

WEBVTT

NOTE duration: "02:28:08.875"

NOTE Confidence: 0.87231445

00:00:00.320 --> 00:00:01.139 Good afternoon,

NOTE Confidence: 0.9970703

00:00:02.080 --> 00:00:02.580 everyone.

NOTE Confidence: 0.98168945

00:00:03.360 --> 00:00:04.420 Welcome to

NOTE Confidence: 0.6934204

00:00:05.120 --> 00:00:06.819 Yale pathology ground runs.

NOTE Confidence: 0.99731445

00:00:08.720 --> 00:00:10.500 Have you ever wondered

NOTE Confidence: 0.95820755

00:00:11.039 --> 00:00:13.059 why children get a malignant

NOTE Confidence: 0.95820755

00:00:13.119 --> 00:00:14.480 tumor at such a young

NOTE Confidence: 0.95820755

00:00:14.480 --> 00:00:14.980 age?

NOTE Confidence: 0.9797058

00:00:15.415 --> 00:00:16.775 Today, you're going to hear

NOTE Confidence: 0.9797058

00:00:16.775 --> 00:00:17.675 from the expert,

NOTE Confidence: 0.947168

00:00:18.055 --> 00:00:19.915 get some answers from him.

NOTE Confidence: 0.90898436

00:00:20.855 --> 00:00:23.275 So it's my great honor

NOTE Confidence: 0.96276855

00:00:23.575 --> 00:00:25.275 and pleasure to introduce

NOTE Confidence: 0.65301514

00:00:25.654 --> 00:00:27.275 today's Guam Run speaker,

NOTE Confidence: 0.5786133

00:00:27.895 --> 00:00:28.395 doctor
NOTE Confidence: 0.98657227
00:00:28.935 --> 00:00:30.154 Miguel Reyes.
NOTE Confidence: 0.89212036
00:00:31.439 --> 00:00:33.360 So probably doctor Reyes don't
NOTE Confidence: 0.89212036
00:00:33.360 --> 00:00:34.500 need a introduction.
NOTE Confidence: 0.8326823
00:00:35.440 --> 00:00:35.940 He's,
NOTE Confidence: 0.9589656
00:00:36.559 --> 00:00:38.479 you know, has friends here
NOTE Confidence: 0.9589656
00:00:38.479 --> 00:00:40.420 with Yale for many years,
NOTE Confidence: 0.9589656
00:00:40.479 --> 00:00:41.460 has a great
NOTE Confidence: 0.795166
00:00:41.920 --> 00:00:42.420 friend.
NOTE Confidence: 0.9926758
00:00:43.040 --> 00:00:44.260 So just briefly,
NOTE Confidence: 0.95751953
00:00:45.055 --> 00:00:47.455 doctor Reyes is currently a
NOTE Confidence: 0.95751953
00:00:47.455 --> 00:00:48.675 clinical professor
NOTE Confidence: 0.65649414
00:00:49.534 --> 00:00:51.155 and pediatric and
NOTE Confidence: 0.5209961
00:00:51.455 --> 00:00:51.955 developmental
NOTE Confidence: 0.9550781
00:00:52.335 --> 00:00:53.955 pathology division chief
NOTE Confidence: 0.9032593
00:00:54.495 --> 00:00:55.955 in the department of pathology

NOTE Confidence: 0.9032593
00:00:56.015 --> 00:00:57.395 and laboratory medicine
NOTE Confidence: 0.99902344
00:00:58.175 --> 00:00:58.675 at
NOTE Confidence: 0.93880206
00:01:00.230 --> 00:01:01.450 University of Miami,
NOTE Confidence: 0.9403076
00:01:02.950 --> 00:01:04.330 Miller School of Medicine.
NOTE Confidence: 0.7722168
00:01:05.830 --> 00:01:07.050 Doctor. Reyes
NOTE Confidence: 0.98657227
00:01:07.590 --> 00:01:09.530 completed his medical education
NOTE Confidence: 0.9614258
00:01:10.150 --> 00:01:10.970 in National
NOTE Confidence: 0.99853516
00:01:11.590 --> 00:01:12.090 Autonomous
NOTE Confidence: 1
00:01:13.030 --> 00:01:13.530 University
NOTE Confidence: 0.9995117
00:01:14.229 --> 00:01:14.970 of Mexico
NOTE Confidence: 0.86271155
00:01:15.615 --> 00:01:16.355 in 1980s,
NOTE Confidence: 0.99853516
00:01:18.334 --> 00:01:19.155 followed by
NOTE Confidence: 0.8667806
00:01:19.455 --> 00:01:21.955 residency training and in Anatomic
NOTE Confidence: 0.8667806
00:01:22.095 --> 00:01:22.595 Passage
NOTE Confidence: 0.9875488
00:01:23.055 --> 00:01:23.555 there.
NOTE Confidence: 0.9983724

00:01:24.655 --> 00:01:25.875 After several years
NOTE Confidence: 0.97537667

00:01:26.334 --> 00:01:28.735 working as attending physician in
NOTE Confidence: 0.97537667

00:01:28.735 --> 00:01:29.795 National Institute
NOTE Confidence: 0.9760742

00:01:30.509 --> 00:01:31.250 of Pediatrics,
NOTE Confidence: 0.8579915

00:01:32.189 --> 00:01:33.950 he decided come to United
NOTE Confidence: 0.8579915

00:01:33.950 --> 00:01:34.450 States
NOTE Confidence: 0.8140869

00:01:34.990 --> 00:01:37.009 and to continue his medical
NOTE Confidence: 0.8140869

00:01:37.149 --> 00:01:37.649 careers.
NOTE Confidence: 0.9348755

00:01:39.229 --> 00:01:41.090 Doctor. Reyes got his pediatric
NOTE Confidence: 0.9348755

00:01:41.149 --> 00:01:42.770 pathology fellowship training
NOTE Confidence: 0.9847412

00:01:43.149 --> 00:01:45.009 in nineteen ninety four
NOTE Confidence: 0.98579913

00:01:45.765 --> 00:01:48.245 from Children's Memorial Hospital of
NOTE Confidence: 0.98579913

00:01:48.245 --> 00:01:48.745 Northwestern
NOTE Confidence: 0.7265625

00:01:49.285 --> 00:01:49.785 University.
NOTE Confidence: 0.9667969

00:01:51.365 --> 00:01:52.425 After the fellowship,
NOTE Confidence: 0.99902344

00:01:53.285 --> 00:01:54.745 he joined Yale

NOTE Confidence: 0.99790734
00:01:55.205 --> 00:01:57.205 as a faculty member in
NOTE Confidence: 0.99790734
00:01:57.205 --> 00:01:58.265 our department
NOTE Confidence: 0.77246094
00:01:59.365 --> 00:01:59.865 and
NOTE Confidence: 0.8313802
00:02:01.180 --> 00:02:02.640 raised his rank
NOTE Confidence: 0.9523112
00:02:03.180 --> 00:02:05.260 from assistant professor to full
NOTE Confidence: 0.9523112
00:02:05.260 --> 00:02:05.760 professors.
NOTE Confidence: 0.9742187
00:02:07.979 --> 00:02:10.639 So after spending fourteen years
NOTE Confidence: 1
00:02:10.940 --> 00:02:11.440 in
NOTE Confidence: 0.99609375
00:02:11.900 --> 00:02:12.400 Yale,
NOTE Confidence: 0.9995117
00:02:13.100 --> 00:02:14.400 he relocated
NOTE Confidence: 0.99560547
00:02:14.700 --> 00:02:15.600 to Pittsburgh
NOTE Confidence: 0.9560547
00:02:16.935 --> 00:02:18.955 to become the chief pathologist
NOTE Confidence: 0.9440918
00:02:19.255 --> 00:02:21.595 in Children's Hospital of Pittsburgh,
NOTE Confidence: 0.68237305
00:02:22.455 --> 00:02:22.955 University,
NOTE Confidence: 0.984375
00:02:23.895 --> 00:02:24.395 Pittsburgh
NOTE Confidence: 0.9550781

00:02:25.815 --> 00:02:27.835 until twenty twenty four.
NOTE Confidence: 0.85429686

00:02:29.419 --> 00:02:31.840 Doctor. Reyes is world renowned
NOTE Confidence: 0.9523926

00:02:32.620 --> 00:02:33.760 pediatric pathologist,
NOTE Confidence: 0.98291016

00:02:34.620 --> 00:02:35.120 has
NOTE Confidence: 0.8307292

00:02:35.419 --> 00:02:37.280 make significant contribution
NOTE Confidence: 0.99902344

00:02:38.060 --> 00:02:39.040 to the pediatric
NOTE Confidence: 0.803833

00:02:39.340 --> 00:02:40.480 pathologist communities,
NOTE Confidence: 0.9978841

00:02:41.180 --> 00:02:42.880 including clinical practice,
NOTE Confidence: 1

00:02:43.944 --> 00:02:44.444 translational
NOTE Confidence: 0.8256836

00:02:44.745 --> 00:02:45.245 research,
NOTE Confidence: 1

00:02:45.865 --> 00:02:46.365 and
NOTE Confidence: 0.9740397

00:02:46.825 --> 00:02:48.044 medical educations.
NOTE Confidence: 0.9611468

00:02:50.425 --> 00:02:52.925 He was the past president
NOTE Confidence: 0.9611468

00:02:53.145 --> 00:02:54.125 of International
NOTE Confidence: 0.9468994

00:02:54.425 --> 00:02:55.565 Pediatric Massage
NOTE Confidence: 0.99609375

00:02:55.944 --> 00:02:56.444 Associations

NOTE Confidence: 0.84488934
00:02:57.700 --> 00:02:58.519 and the Society
NOTE Confidence: 0.98944765
00:02:59.780 --> 00:03:01.239 of Pediatric Massage.
NOTE Confidence: 0.54974365
00:03:03.139 --> 00:03:04.120 Doctor. Reyes
NOTE Confidence: 0.83935547
00:03:04.659 --> 00:03:05.159 published
NOTE Confidence: 0.95214844
00:03:05.859 --> 00:03:07.780 more than three hundred peer
NOTE Confidence: 0.95214844
00:03:07.780 --> 00:03:08.840 review articles,
NOTE Confidence: 0.5019531
00:03:09.859 --> 00:03:10.359 book
NOTE Confidence: 0.8317261
00:03:10.819 --> 00:03:12.519 books and book chapters.
NOTE Confidence: 0.96516925
00:03:15.514 --> 00:03:17.375 So he is the standing
NOTE Confidence: 0.96516925
00:03:17.595 --> 00:03:18.095 editors
NOTE Confidence: 0.93725586
00:03:18.635 --> 00:03:19.935 of the WHO classification
NOTE Confidence: 0.9707031
00:03:20.794 --> 00:03:21.375 of tumors
NOTE Confidence: 0.78674316
00:03:22.155 --> 00:03:23.375 editorial board.
NOTE Confidence: 0.6055908
00:03:23.834 --> 00:03:24.814 And he was the
NOTE Confidence: 0.99902344
00:03:25.595 --> 00:03:26.095 expert
NOTE Confidence: 0.99609375

00:03:26.474 --> 00:03:26.974 editor
NOTE Confidence: 0.9857585
00:03:27.900 --> 00:03:30.060 for the newly released WHO
NOTE Confidence: 0.9857585
00:03:30.060 --> 00:03:30.560 classification
NOTE Confidence: 0.99316406
00:03:31.020 --> 00:03:31.760 of tumor
NOTE Confidence: 0.6425781
00:03:32.540 --> 00:03:33.040 pediatric
NOTE Confidence: 0.99853516
00:03:33.980 --> 00:03:34.480 tumors.
NOTE Confidence: 0.8977051
00:03:35.660 --> 00:03:37.360 So although, you know,
NOTE Confidence: 0.9814453
00:03:37.740 --> 00:03:39.360 the current WHO classification
NOTE Confidence: 0.9970703
00:03:39.740 --> 00:03:40.240 tumor
NOTE Confidence: 0.9628906
00:03:40.780 --> 00:03:41.900 already in his
NOTE Confidence: 0.84716797
00:03:42.515 --> 00:03:43.015 its
NOTE Confidence: 0.8619385
00:03:43.315 --> 00:03:44.455 fifth edition.
NOTE Confidence: 0.95133466
00:03:45.474 --> 00:03:46.855 The pediatric tumors
NOTE Confidence: 0.98535156
00:03:47.555 --> 00:03:48.855 is the first time
NOTE Confidence: 0.9479167
00:03:49.155 --> 00:03:50.775 WHO has including
NOTE Confidence: 0.9638672
00:03:51.075 --> 00:03:52.455 a separate volume

NOTE Confidence: 0.98791504
00:03:53.235 --> 00:03:54.935 on the pediatric tumors.
NOTE Confidence: 0.92094725
00:03:56.720 --> 00:03:58.660 There's no doubt this publication
NOTE Confidence: 0.98811847
00:03:59.200 --> 00:04:00.420 will help improving
NOTE Confidence: 0.9958496
00:04:01.280 --> 00:04:02.099 the standardization
NOTE Confidence: 0.9995117
00:04:02.800 --> 00:04:03.620 of pediatric
NOTE Confidence: 0.9980469
00:04:04.080 --> 00:04:05.300 tumor diagnosis
NOTE Confidence: 0.9797363
00:04:06.000 --> 00:04:06.739 and facilitate
NOTE Confidence: 0.7470703
00:04:07.760 --> 00:04:08.580 the translate
NOTE Confidence: 0.99934894
00:04:09.615 --> 00:04:11.155 translation of diagnostic
NOTE Confidence: 0.9980469
00:04:11.455 --> 00:04:12.675 research into
NOTE Confidence: 1
00:04:13.215 --> 00:04:13.715 practice
NOTE Confidence: 0.99560547
00:04:14.255 --> 00:04:14.755 worldwide.
NOTE Confidence: 0.9800415
00:04:15.855 --> 00:04:17.795 So without further ado,
NOTE Confidence: 0.8501465
00:04:18.495 --> 00:04:20.835 please join me, Wolfgang. Reyes.
NOTE Confidence: 0.9980469
00:04:28.350 --> 00:04:29.810 Well, thank you very much
NOTE Confidence: 0.8508301

00:04:30.670 --> 00:04:31.410 for being
NOTE Confidence: 0.8059082

00:04:32.190 --> 00:04:33.570 first for inviting me,
NOTE Confidence: 0.92871094

00:04:34.110 --> 00:04:37.010 then for, presenting me as
NOTE Confidence: 0.92871094

00:04:37.150 --> 00:04:38.770 in such a kindly manner.
NOTE Confidence: 0.9694824

00:04:40.214 --> 00:04:42.295 I recognize and have a
NOTE Confidence: 0.9694824

00:04:42.295 --> 00:04:42.775 lot of,
NOTE Confidence: 0.98535156

00:04:44.135 --> 00:04:44.635 fun
NOTE Confidence: 0.99751204

00:04:45.415 --> 00:04:46.615 to see many of my
NOTE Confidence: 0.99751204

00:04:46.615 --> 00:04:47.435 former trainees,
NOTE Confidence: 0.99609375

00:04:48.214 --> 00:04:48.714 colleagues,
NOTE Confidence: 0.97387695

00:04:49.095 --> 00:04:49.595 friends,
NOTE Confidence: 0.9996745

00:04:50.935 --> 00:04:52.235 and people that
NOTE Confidence: 0.97194415

00:04:52.630 --> 00:04:54.470 taught me how to become
NOTE Confidence: 0.97194415

00:04:54.470 --> 00:04:55.770 an academic pathologist.
NOTE Confidence: 0.7566406

00:04:56.470 --> 00:04:58.890 There's Costa, doctor Morrow here,
NOTE Confidence: 0.99121094

00:04:59.510 --> 00:05:00.010 and

NOTE Confidence: 0.72424316
00:05:00.630 --> 00:05:01.690 David here.
NOTE Confidence: 0.98999023
00:05:02.310 --> 00:05:03.050 We interacted
NOTE Confidence: 0.91048175
00:05:03.510 --> 00:05:04.710 the other day with here.
NOTE Confidence: 0.91048175
00:05:04.710 --> 00:05:06.550 We interacted with them many
NOTE Confidence: 0.91048175
00:05:06.550 --> 00:05:07.190 years, and,
NOTE Confidence: 0.97836304
00:05:08.225 --> 00:05:09.185 it has been an honor
NOTE Confidence: 0.97836304
00:05:09.185 --> 00:05:10.145 for me. This is really
NOTE Confidence: 0.97836304
00:05:10.145 --> 00:05:11.745 my academic home, so I
NOTE Confidence: 0.97836304
00:05:11.745 --> 00:05:13.605 am very glad to return.
NOTE Confidence: 0.98431396
00:05:15.505 --> 00:05:17.904 Today, I will present something
NOTE Confidence: 0.98431396
00:05:17.904 --> 00:05:19.585 that is different from, the
NOTE Confidence: 0.98431396
00:05:19.585 --> 00:05:21.285 previous conference that I presented
NOTE Confidence: 0.98431396
00:05:21.345 --> 00:05:23.319 here about eight years ago.
NOTE Confidence: 0.96069336
00:05:23.699 --> 00:05:25.620 Today, we will, talk about
NOTE Confidence: 0.96069336
00:05:25.620 --> 00:05:26.120 how
NOTE Confidence: 0.97216797

00:05:26.500 --> 00:05:28.099 looking at the slice in
NOTE Confidence: 0.97216797

00:05:28.099 --> 00:05:29.240 pediatric lesions
NOTE Confidence: 0.99902344

00:05:29.699 --> 00:05:31.080 can allow you to identify
NOTE Confidence: 0.9951172

00:05:32.419 --> 00:05:33.240 cancer predisposition
NOTE Confidence: 0.9907532

00:05:33.620 --> 00:05:35.379 syndromes. This is a topic
NOTE Confidence: 0.9907532

00:05:35.379 --> 00:05:35.940 that I have,
NOTE Confidence: 0.8701172

00:05:37.645 --> 00:05:38.145 cultivated,
NOTE Confidence: 0.99727005

00:05:39.005 --> 00:05:40.685 extensively in the last few
NOTE Confidence: 0.99727005

00:05:40.685 --> 00:05:42.625 years. How do I advance
NOTE Confidence: 0.99727005

00:05:42.685 --> 00:05:43.185 this?
NOTE Confidence: 0.9660034

00:05:49.420 --> 00:05:50.779 Oh, thank you very much.
NOTE Confidence: 0.9660034

00:05:50.779 --> 00:05:51.920 Didn't see that.
NOTE Confidence: 0.9893799

00:05:52.860 --> 00:05:54.560 So let's move on.
NOTE Confidence: 0.93847656

00:05:56.779 --> 00:05:58.240 I have nothing to declare,
NOTE Confidence: 0.9638184

00:05:59.420 --> 00:06:01.200 and, this is the outline,
NOTE Confidence: 0.9945243

00:06:02.545 --> 00:06:04.065 and we will go directly

NOTE Confidence: 0.9945243
00:06:04.065 --> 00:06:04.805 into matters.
NOTE Confidence: 0.9461806
00:06:05.825 --> 00:06:07.105 At the beginning of last
NOTE Confidence: 0.9461806
00:06:07.105 --> 00:06:09.125 year, the official's archive
NOTE Confidence: 0.99506295
00:06:09.425 --> 00:06:11.025 journal, the oldest journal of
NOTE Confidence: 0.99506295
00:06:11.025 --> 00:06:12.485 pathology in the world,
NOTE Confidence: 0.93621534
00:06:13.265 --> 00:06:15.265 published a special issue in
NOTE Confidence: 0.93621534
00:06:15.265 --> 00:06:17.180 which, my colleague from Madrid,
NOTE Confidence: 0.93621534
00:06:17.180 --> 00:06:19.100 Doctor. Isabel Colmenero, the chief
NOTE Confidence: 0.93621534
00:06:19.100 --> 00:06:21.180 pathologist at Hospital del Nino
NOTE Confidence: 0.93621534
00:06:21.180 --> 00:06:21.680 Jesus
NOTE Confidence: 0.90356445
00:06:21.980 --> 00:06:22.720 in Madrid,
NOTE Confidence: 0.9986049
00:06:23.580 --> 00:06:25.020 and I put together a
NOTE Confidence: 0.9986049
00:06:25.020 --> 00:06:26.000 collection of
NOTE Confidence: 0.94998604
00:06:26.460 --> 00:06:27.980 twelve papers, ten of twelve
NOTE Confidence: 0.94998604
00:06:27.980 --> 00:06:30.135 papers, and an editorial. We
NOTE Confidence: 0.94998604

00:06:30.135 --> 00:06:31.495 invited all the authors that
NOTE Confidence: 0.94998604

00:06:31.495 --> 00:06:33.654 contributed to this special issue
NOTE Confidence: 0.94998604

00:06:33.654 --> 00:06:34.154 highlighting
NOTE Confidence: 0.9382324

00:06:34.775 --> 00:06:35.175 the,
NOTE Confidence: 0.9959717

00:06:35.575 --> 00:06:37.595 challenges and novelties in pediatric
NOTE Confidence: 0.9959717

00:06:37.654 --> 00:06:38.154 pathology.
NOTE Confidence: 0.973114

00:06:38.535 --> 00:06:39.735 I invite you to review
NOTE Confidence: 0.973114

00:06:39.735 --> 00:06:41.095 the special issue because it
NOTE Confidence: 0.973114

00:06:41.095 --> 00:06:43.415 contains interesting information that is
NOTE Confidence: 0.973114

00:06:43.415 --> 00:06:43.915 mostly
NOTE Confidence: 0.9886719

00:06:44.630 --> 00:06:46.889 missed by the adult pathology,
NOTE Confidence: 0.919637

00:06:47.910 --> 00:06:49.770 sphere, the adult adult pathology
NOTE Confidence: 0.919637

00:06:49.990 --> 00:06:50.490 circles.
NOTE Confidence: 0.9897461

00:06:51.589 --> 00:06:52.330 In this
NOTE Confidence: 0.7663574

00:06:53.669 --> 00:06:54.169 issue,
NOTE Confidence: 0.99975586

00:06:54.470 --> 00:06:55.529 one of the papers

NOTE Confidence: 0.9967913

00:06:55.990 --> 00:06:57.830 was specifically dedicated to the

NOTE Confidence: 0.9967913

00:06:57.830 --> 00:06:58.889 pediatric cancer

NOTE Confidence: 0.9487467

00:06:59.214 --> 00:07:01.634 position syndromes in the, outside,

NOTE Confidence: 0.9638184

00:07:02.095 --> 00:07:04.035 of the central nervous system.

NOTE Confidence: 0.9888071

00:07:04.495 --> 00:07:06.014 And, I also invite you

NOTE Confidence: 0.9888071

00:07:06.014 --> 00:07:07.294 to read this paper because

NOTE Confidence: 0.9888071

00:07:07.294 --> 00:07:08.275 this will summarize

NOTE Confidence: 0.9994575

00:07:08.654 --> 00:07:09.615 what I am going to

NOTE Confidence: 0.9994575

00:07:09.615 --> 00:07:10.914 present to you today.

NOTE Confidence: 0.94763184

00:07:13.940 --> 00:07:15.320 Okay. It's not advancing.

NOTE Confidence: 0.9680542

00:07:15.699 --> 00:07:17.460 Okay. Here. So how do

NOTE Confidence: 0.9680542

00:07:17.460 --> 00:07:19.800 we define cancer predisposition syndromes?

NOTE Confidence: 0.99386936

00:07:20.740 --> 00:07:22.979 Heritable condition that increases the

NOTE Confidence: 0.99386936

00:07:22.979 --> 00:07:24.820 child's risk of developing cancer

NOTE Confidence: 0.99386936

00:07:24.820 --> 00:07:26.580 due to general mutations in

NOTE Confidence: 0.99386936

00:07:26.580 --> 00:07:27.639 a gene regulating
NOTE Confidence: 0.93223065

00:07:28.235 --> 00:07:29.995 cell growth, DNA repair, or
NOTE Confidence: 0.93223065

00:07:29.995 --> 00:07:31.935 genomic integrity. That's in general
NOTE Confidence: 0.93223065

00:07:31.995 --> 00:07:33.755 where a cancer predisposition syndrome
NOTE Confidence: 0.93223065

00:07:33.755 --> 00:07:34.875 is. And, of course, this
NOTE Confidence: 0.93223065

00:07:34.875 --> 00:07:36.175 affects not only children,
NOTE Confidence: 0.87332153

00:07:36.475 --> 00:07:38.955 but, adults too, but adults
NOTE Confidence: 0.87332153

00:07:38.955 --> 00:07:40.955 affected with these conditions bring
NOTE Confidence: 0.87332153

00:07:40.955 --> 00:07:42.794 these cons conditions since they
NOTE Confidence: 0.87332153

00:07:42.794 --> 00:07:43.110 are
NOTE Confidence: 0.90239257

00:07:43.990 --> 00:07:46.090 children. So we pediatric pathologists
NOTE Confidence: 0.98860675

00:07:46.550 --> 00:07:48.469 are at the specific point
NOTE Confidence: 0.98860675

00:07:48.469 --> 00:07:50.569 to identify these entities
NOTE Confidence: 0.9769694

00:07:50.870 --> 00:07:53.050 early, early enough to intervene
NOTE Confidence: 0.9769694

00:07:53.349 --> 00:07:55.349 and modify the outcomes of
NOTE Confidence: 0.9769694

00:07:55.349 --> 00:07:56.250 these patients.

NOTE Confidence: 0.93033856

00:07:56.915 --> 00:07:58.375 They are, representing,

NOTE Confidence: 0.6621094

00:08:00.195 --> 00:08:00.935 in inherited

NOTE Confidence: 0.8418213

00:08:01.235 --> 00:08:04.375 risk, usually autosomal dominant, occasionally,

NOTE Confidence: 0.536499

00:08:04.835 --> 00:08:05.495 the novel,

NOTE Confidence: 0.98266006

00:08:05.795 --> 00:08:07.875 and the tumor often appears

NOTE Confidence: 0.98266006

00:08:07.875 --> 00:08:09.715 at younger ages than, general

NOTE Confidence: 0.98266006

00:08:09.715 --> 00:08:11.155 population. The same type of

NOTE Confidence: 0.98266006

00:08:11.155 --> 00:08:12.115 tumor that you see in

NOTE Confidence: 0.98266006

00:08:12.115 --> 00:08:13.530 adults when it appears in

NOTE Confidence: 0.98266006

00:08:13.530 --> 00:08:14.810 a child, it may be

NOTE Confidence: 0.98266006

00:08:14.810 --> 00:08:16.250 the first clue for you

NOTE Confidence: 0.98266006

00:08:16.250 --> 00:08:17.610 to look for a cancer

NOTE Confidence: 0.98266006

00:08:17.610 --> 00:08:18.110 predisposition

NOTE Confidence: 0.9995117

00:08:18.490 --> 00:08:18.990 syndrome.

NOTE Confidence: 0.9695638

00:08:19.690 --> 00:08:21.530 Multiple of these, many of

NOTE Confidence: 0.9695638

00:08:21.530 --> 00:08:23.070 these tumors are rare
NOTE Confidence: 0.98157555

00:08:23.450 --> 00:08:25.710 and, children with multiple primaries
NOTE Confidence: 0.98157555

00:08:25.850 --> 00:08:28.324 or unusual types are specifically
NOTE Confidence: 0.98157555

00:08:28.384 --> 00:08:29.764 a red flag to identify.
NOTE Confidence: 0.9760742

00:08:30.384 --> 00:08:32.065 The family history may show
NOTE Confidence: 0.9760742

00:08:32.065 --> 00:08:34.225 clustering of specific or related
NOTE Confidence: 0.9760742

00:08:34.225 --> 00:08:36.325 causes, but sometimes it doesn't.
NOTE Confidence: 0.9786266

00:08:37.105 --> 00:08:39.125 Approximately ten percent of pediatric
NOTE Confidence: 0.9786266

00:08:39.184 --> 00:08:40.885 cancers are due to germline
NOTE Confidence: 0.9786266

00:08:41.024 --> 00:08:41.524 alterations
NOTE Confidence: 0.9470215

00:08:42.380 --> 00:08:45.020 and early frequent recognition is
NOTE Confidence: 0.9470215

00:08:45.020 --> 00:08:45.679 a key,
NOTE Confidence: 0.850708

00:08:47.980 --> 00:08:49.520 to, provide adequate surveillance
NOTE Confidence: 0.8773804

00:08:49.900 --> 00:08:51.840 therapy and family assessment.
NOTE Confidence: 0.95410156

00:08:52.540 --> 00:08:53.040 So
NOTE Confidence: 0.9501926

00:08:53.340 --> 00:08:54.700 the point that I will

NOTE Confidence: 0.9501926
00:08:54.700 --> 00:08:56.505 emphasize today is that frequently
NOTE Confidence: 0.9501926
00:08:56.905 --> 00:08:59.545 histology provides the first clue,
NOTE Confidence: 0.9501926
00:08:59.545 --> 00:09:00.345 but you just have to
NOTE Confidence: 0.9501926
00:09:00.345 --> 00:09:01.945 keep this in mind. If
NOTE Confidence: 0.9501926
00:09:01.945 --> 00:09:03.804 you don't, you'll miss it.
NOTE Confidence: 0.9951172
00:09:05.465 --> 00:09:05.965 Okay.
NOTE Confidence: 0.97843426
00:09:09.360 --> 00:09:11.120 The pediatric pathologist is then
NOTE Confidence: 0.97843426
00:09:11.120 --> 00:09:12.960 at the crossroads between what
NOTE Confidence: 0.97843426
00:09:12.960 --> 00:09:14.480 is the clinical observation of
NOTE Confidence: 0.97843426
00:09:14.480 --> 00:09:16.240 a patient, the genetics of
NOTE Confidence: 0.97843426
00:09:16.240 --> 00:09:17.620 this patient, and evaluating,
NOTE Confidence: 0.95807755
00:09:18.320 --> 00:09:20.000 the lesions that are under
NOTE Confidence: 0.95807755
00:09:20.000 --> 00:09:20.660 the microscope
NOTE Confidence: 0.97813195
00:09:21.135 --> 00:09:22.595 with the risk of increasing
NOTE Confidence: 0.97813195
00:09:22.815 --> 00:09:23.875 developing cancer.
NOTE Confidence: 0.9885254

00:09:24.735 --> 00:09:25.475 In addition
NOTE Confidence: 0.9788574

00:09:25.855 --> 00:09:27.955 to accurately diagnose these entities,
NOTE Confidence: 0.9989258

00:09:28.335 --> 00:09:29.795 we need to be familiarized
NOTE Confidence: 0.9776367

00:09:30.335 --> 00:09:32.195 with the implications of additional,
NOTE Confidence: 1

00:09:33.535 --> 00:09:34.035 seemingly
NOTE Confidence: 0.93033856

00:09:35.140 --> 00:09:35.640 uninteresting,
NOTE Confidence: 0.9227905

00:09:36.100 --> 00:09:37.779 unimportant findings that I will
NOTE Confidence: 0.9227905

00:09:37.779 --> 00:09:38.600 show you today.
NOTE Confidence: 0.9576823

00:09:38.980 --> 00:09:40.920 And clinical and or laboratory
NOTE Confidence: 0.9576823

00:09:40.980 --> 00:09:41.480 findings,
NOTE Confidence: 0.89853513

00:09:42.420 --> 00:09:44.580 that are apparently minor may
NOTE Confidence: 0.89853513

00:09:44.580 --> 00:09:47.080 be of various important significance.
NOTE Confidence: 0.9549735

00:09:48.014 --> 00:09:48.894 This is taken from the
NOTE Confidence: 0.9549735

00:09:48.894 --> 00:09:50.415 paper of the officials archive
NOTE Confidence: 0.9549735

00:09:50.415 --> 00:09:51.774 journal that I mentioned to
NOTE Confidence: 0.9549735

00:09:51.774 --> 00:09:53.214 you. I'm not going to

NOTE Confidence: 0.9549735

00:09:53.214 --> 00:09:54.894 go over every one of

NOTE Confidence: 0.9549735

00:09:54.894 --> 00:09:56.254 these entities. This is just

NOTE Confidence: 0.9549735

00:09:56.254 --> 00:09:58.175 to emphasize that I I

NOTE Confidence: 0.9549735

00:09:58.175 --> 00:09:59.855 would like you to, review

NOTE Confidence: 0.9549735

00:09:59.855 --> 00:10:01.375 the paper, read it, and

NOTE Confidence: 0.9549735

00:10:01.375 --> 00:10:03.090 and, send us any,

NOTE Confidence: 0.9407756

00:10:03.490 --> 00:10:05.809 comments, criticisms, etcetera. There are

NOTE Confidence: 0.9407756

00:10:05.809 --> 00:10:06.309 many,

NOTE Confidence: 0.9683838

00:10:06.930 --> 00:10:07.750 cancer predisposition

NOTE Confidence: 0.99675566

00:10:08.050 --> 00:10:09.970 syndromes listed here. Obviously, this

NOTE Confidence: 0.99675566

00:10:09.970 --> 00:10:11.090 is just to show you

NOTE Confidence: 0.99675566

00:10:11.090 --> 00:10:12.470 that the paper is long

NOTE Confidence: 0.96606445

00:10:12.850 --> 00:10:13.170 and,

NOTE Confidence: 0.97631836

00:10:13.970 --> 00:10:14.790 full of,

NOTE Confidence: 1

00:10:15.250 --> 00:10:15.750 information

NOTE Confidence: 0.98272705

00:10:16.054 --> 00:10:16.934 that I am not going
NOTE Confidence: 0.98272705

00:10:16.934 --> 00:10:18.214 to be able to detail
NOTE Confidence: 0.98272705

00:10:18.214 --> 00:10:19.654 here. But we are going
NOTE Confidence: 0.98272705

00:10:19.654 --> 00:10:20.054 to,
NOTE Confidence: 0.9937221

00:10:20.855 --> 00:10:22.054 start with one of the
NOTE Confidence: 0.9937221

00:10:22.054 --> 00:10:22.795 most common,
NOTE Confidence: 0.99991864

00:10:23.415 --> 00:10:25.175 pediatric tumors that we see
NOTE Confidence: 0.99991864

00:10:25.175 --> 00:10:25.675 here.
NOTE Confidence: 0.90108234

00:10:26.774 --> 00:10:28.295 This is Wilms' tumor also
NOTE Confidence: 0.90108234

00:10:28.295 --> 00:10:29.274 known as nephroblastoma.
NOTE Confidence: 0.95047

00:10:30.449 --> 00:10:32.290 Nobody really knows what doctor
NOTE Confidence: 0.95047

00:10:32.290 --> 00:10:33.589 Max Williams described.
NOTE Confidence: 0.96203035

00:10:34.529 --> 00:10:36.529 Bruce Beckwith, the late Bruce
NOTE Confidence: 0.96203035

00:10:36.529 --> 00:10:37.809 Beckwith, a good friend of
NOTE Confidence: 0.96203035

00:10:37.809 --> 00:10:39.189 mine, who was the,
NOTE Confidence: 0.9814453

00:10:39.889 --> 00:10:41.569 official reviewer for the children's

NOTE Confidence: 0.9814453
00:10:41.569 --> 00:10:43.429 oncology group for many years,
NOTE Confidence: 0.9461723
00:10:44.365 --> 00:10:46.125 told me several times that
NOTE Confidence: 0.9461723
00:10:46.125 --> 00:10:47.804 nobody really understood what Max
NOTE Confidence: 0.9461723
00:10:47.804 --> 00:10:49.485 Williams, described, but this is
NOTE Confidence: 0.9461723
00:10:49.485 --> 00:10:50.865 the entity that we recognize
NOTE Confidence: 0.9461723
00:10:50.925 --> 00:10:51.904 as Wim's tumor.
NOTE Confidence: 0.9284668
00:10:52.365 --> 00:10:53.804 A tumor that produces a
NOTE Confidence: 0.9284668
00:10:53.804 --> 00:10:54.304 massive
NOTE Confidence: 0.60595703
00:10:54.765 --> 00:10:55.265 growth
NOTE Confidence: 0.9635376
00:10:55.770 --> 00:10:57.530 inside the kidney with a
NOTE Confidence: 0.9635376
00:10:57.530 --> 00:10:59.390 claw sign on the residual
NOTE Confidence: 0.9635376
00:10:59.530 --> 00:11:01.130 kidney that surrounds it as
NOTE Confidence: 0.9635376
00:11:01.130 --> 00:11:02.270 you can see in this,
NOTE Confidence: 0.9180298
00:11:02.730 --> 00:11:04.250 CT scan and this is
NOTE Confidence: 0.9180298
00:11:04.250 --> 00:11:05.630 a classic microscopic
NOTE Confidence: 0.93199325

00:11:06.010 --> 00:11:07.370 appearance. I don't use the
NOTE Confidence: 0.93199325

00:11:07.370 --> 00:11:08.990 gross the gross term
NOTE Confidence: 0.95214844

00:11:09.425 --> 00:11:10.704 because it grosses me out.
NOTE Confidence: 0.95214844

00:11:10.704 --> 00:11:11.684 It's recommending
NOTE Confidence: 0.94249135

00:11:12.064 --> 00:11:13.665 it's it's recommended to get
NOTE Confidence: 0.94249135

00:11:13.665 --> 00:11:14.944 rid of the grossing and
NOTE Confidence: 0.94249135

00:11:14.944 --> 00:11:16.884 gross. But, anyway, the macroscopic
NOTE Confidence: 0.97684735

00:11:17.184 --> 00:11:18.944 appearance of this tumor is
NOTE Confidence: 0.97684735

00:11:18.944 --> 00:11:19.425 this,
NOTE Confidence: 0.97477216

00:11:20.464 --> 00:11:20.964 fleshy,
NOTE Confidence: 0.9109701

00:11:22.120 --> 00:11:25.179 relatively homogeneous, sometimes vocally hem-
orrhagic,
NOTE Confidence: 0.97265625

00:11:25.880 --> 00:11:27.880 sometimes very hemorrhagic. And then
NOTE Confidence: 0.97265625

00:11:27.880 --> 00:11:28.920 you can appreciate that there
NOTE Confidence: 0.97265625

00:11:28.920 --> 00:11:30.600 is a little rim of
NOTE Confidence: 0.97265625

00:11:30.600 --> 00:11:31.899 kidney that,
NOTE Confidence: 0.89001465

00:11:33.000 --> 00:11:33.740 is still,
NOTE Confidence: 0.95532733

00:11:34.315 --> 00:11:36.394 preserved there. This is Bruce
NOTE Confidence: 0.95532733

00:11:36.394 --> 00:11:37.995 Beckwith here visiting at Yale.
NOTE Confidence: 0.95532733

00:11:37.995 --> 00:11:38.875 This is me, believe it
NOTE Confidence: 0.95532733

00:11:38.875 --> 00:11:40.554 or not. I still didn't
NOTE Confidence: 0.95532733

00:11:40.554 --> 00:11:42.014 stain my beard white.
NOTE Confidence: 0.97693586

00:11:42.394 --> 00:11:43.834 And, this is a few
NOTE Confidence: 0.97693586

00:11:43.834 --> 00:11:45.195 years later, ten years later,
NOTE Confidence: 0.97693586

00:11:45.195 --> 00:11:46.315 when we met together in
NOTE Confidence: 0.97693586

00:11:46.315 --> 00:11:47.535 Tours, France.
NOTE Confidence: 0.77870846

00:11:48.980 --> 00:11:50.920 This Beckwith, William and syndrome
NOTE Confidence: 0.77870846

00:11:51.139 --> 00:11:52.660 and other syndromes such as
NOTE Confidence: 0.77870846

00:11:52.660 --> 00:11:53.980 Wagner, Dennis Trash,
NOTE Confidence: 0.86027014

00:11:54.420 --> 00:11:55.399 Simpson Golami,
NOTE Confidence: 0.8685913

00:11:56.179 --> 00:11:57.399 Behamel, Fraser,
NOTE Confidence: 0.7683309

00:11:57.700 --> 00:11:59.700 Bloom syndrome, Dyson one are

NOTE Confidence: 0.7683309
00:11:59.700 --> 00:12:00.200 associated
NOTE Confidence: 0.935214
00:12:00.579 --> 00:12:02.179 with Wilms' tumors, and we
NOTE Confidence: 0.935214
00:12:02.179 --> 00:12:04.065 will review how they look
NOTE Confidence: 0.935214
00:12:04.065 --> 00:12:04.565 at,
NOTE Confidence: 0.9975586
00:12:04.945 --> 00:12:05.764 very briefly.
NOTE Confidence: 0.93990016
00:12:06.144 --> 00:12:08.485 Triphasic Wilms tumors with epithelial
NOTE Confidence: 0.93990016
00:12:08.704 --> 00:12:11.904 tubular components, blastema components, and
NOTE Confidence: 0.93990016
00:12:11.904 --> 00:12:14.005 stromal components are the rule.
NOTE Confidence: 0.93990016
00:12:14.144 --> 00:12:16.005 This is a blastema predominant
NOTE Confidence: 0.93990016
00:12:16.304 --> 00:12:17.824 component and this is a
NOTE Confidence: 0.93990016
00:12:17.824 --> 00:12:20.220 stromal predominant WIMS tumor. And
NOTE Confidence: 0.93990016
00:12:20.220 --> 00:12:21.980 sometimes you can have very
NOTE Confidence: 0.93990016
00:12:21.980 --> 00:12:24.399 clear muscle skeletal muscle differentiation
NOTE Confidence: 0.93990016
00:12:24.459 --> 00:12:25.660 that you can appreciate in
NOTE Confidence: 0.93990016
00:12:25.660 --> 00:12:27.679 these pictures from WIMS tumors.
NOTE Confidence: 0.93990016

00:12:27.980 --> 00:12:29.100 In the past, we used
NOTE Confidence: 0.93990016

00:12:29.100 --> 00:12:31.260 to call this territory variant
NOTE Confidence: 0.93990016

00:12:31.260 --> 00:12:32.459 of a WIMS tumor when
NOTE Confidence: 0.93990016

00:12:32.459 --> 00:12:33.995 there are components such as
NOTE Confidence: 0.93990016

00:12:33.995 --> 00:12:34.975 skeletal muscle,
NOTE Confidence: 0.8457031

00:12:35.915 --> 00:12:36.415 keratinizing
NOTE Confidence: 0.90785724

00:12:36.795 --> 00:12:39.054 squamous epithelium, and even osteoid
NOTE Confidence: 0.90785724

00:12:39.195 --> 00:12:39.695 formations.
NOTE Confidence: 0.8756348

00:12:41.035 --> 00:12:43.035 The war territory is not,
NOTE Confidence: 0.92492676

00:12:43.835 --> 00:12:44.975 recommended anymore.
NOTE Confidence: 0.9494873

00:12:45.595 --> 00:12:46.875 And after you make the
NOTE Confidence: 0.9494873

00:12:46.875 --> 00:12:48.959 diagnosis so what happens? Doctor,
NOTE Confidence: 0.80737305

00:12:51.260 --> 00:12:52.399 Farzana Pankaj,
NOTE Confidence: 0.9471334

00:12:52.860 --> 00:12:54.459 Pankar comes with you and
NOTE Confidence: 0.9471334

00:12:54.459 --> 00:12:55.519 tells you, Miguel,
NOTE Confidence: 0.78100586

00:12:56.140 --> 00:12:57.420 yesterday there was an an

NOTE Confidence: 0.78100586
00:12:57.420 --> 00:12:58.480 a rectomy specimen.
NOTE Confidence: 0.9490182
00:12:58.860 --> 00:12:59.899 Can you tell us what
NOTE Confidence: 0.9490182
00:12:59.899 --> 00:13:00.779 is it? Is it a
NOTE Confidence: 0.9490182
00:13:00.779 --> 00:13:01.899 Williams tumor? And then the
NOTE Confidence: 0.9490182
00:13:01.899 --> 00:13:03.195 first thing is to confirm.
NOTE Confidence: 0.9490182
00:13:03.355 --> 00:13:04.554 Yes. It's a woman's tumor,
NOTE Confidence: 0.9490182
00:13:04.554 --> 00:13:05.934 Farzana. Right? Good.
NOTE Confidence: 0.9675293
00:13:06.315 --> 00:13:07.215 Next question.
NOTE Confidence: 0.9906006
00:13:07.995 --> 00:13:08.975 What is the stage?
NOTE Confidence: 0.9711914
00:13:09.355 --> 00:13:11.035 Okay. And then is it
NOTE Confidence: 0.9711914
00:13:11.035 --> 00:13:12.335 favorable or unfavorable?
NOTE Confidence: 0.9831543
00:13:12.715 --> 00:13:13.934 And how do you distinguish
NOTE Confidence: 0.9831543
00:13:13.995 --> 00:13:15.515 that? Because that takes makes
NOTE Confidence: 0.9831543
00:13:15.515 --> 00:13:16.895 a difference in the treatment
NOTE Confidence: 0.9831543
00:13:16.955 --> 00:13:17.775 of the patient.
NOTE Confidence: 0.9579468

00:13:18.280 --> 00:13:19.640 And then when I respond
NOTE Confidence: 0.9579468

00:13:19.640 --> 00:13:20.620 these three questions,
NOTE Confidence: 0.9994071

00:13:21.000 --> 00:13:22.440 she goes back to her
NOTE Confidence: 0.9994071

00:13:22.440 --> 00:13:23.720 office to look at the
NOTE Confidence: 0.9994071

00:13:23.720 --> 00:13:25.100 patient and start chemotherapy.
NOTE Confidence: 0.9996745

00:13:25.720 --> 00:13:26.620 Is that correct?
NOTE Confidence: 0.9824219

00:13:28.120 --> 00:13:28.620 However,
NOTE Confidence: 0.9142253

00:13:31.085 --> 00:13:32.865 favorable histology is
NOTE Confidence: 0.98947483

00:13:33.325 --> 00:13:34.785 identified with anaplastic
NOTE Confidence: 0.921875

00:13:35.725 --> 00:13:37.425 mitosis or, hyperchromatic
NOTE Confidence: 0.9622026

00:13:37.805 --> 00:13:39.245 nucleomegaly. You have to have
NOTE Confidence: 0.9622026

00:13:39.245 --> 00:13:40.525 both of them for the
NOTE Confidence: 0.9622026

00:13:40.525 --> 00:13:42.125 diagnosis. I listed here the
NOTE Confidence: 0.9622026

00:13:42.125 --> 00:13:43.405 criteria. I'm not gonna go
NOTE Confidence: 0.9622026

00:13:43.405 --> 00:13:44.145 over them,
NOTE Confidence: 1

00:13:44.525 --> 00:13:45.184 in detail

NOTE Confidence: 0.9716163

00:13:45.670 --> 00:13:47.210 And this is another atypical

NOTE Confidence: 0.9716163

00:13:47.350 --> 00:13:50.010 mitosis and, hyperchromatic nucleomegaly

NOTE Confidence: 0.96595347

00:13:50.390 --> 00:13:51.429 at least three times the

NOTE Confidence: 0.96595347

00:13:51.429 --> 00:13:53.210 size of adjacent non anaplastic

NOTE Confidence: 0.96595347

00:13:53.429 --> 00:13:53.929 nuclei.

NOTE Confidence: 0.96125424

00:13:54.630 --> 00:13:56.570 Immuno histo chemistry is irrelevant

NOTE Confidence: 0.96125424

00:13:56.790 --> 00:13:58.230 in the diagnosis and study

NOTE Confidence: 0.96125424

00:13:58.230 --> 00:13:59.990 of WIMS tumor patients, but

NOTE Confidence: 0.96125424

00:13:59.990 --> 00:14:01.635 I listed here what is

NOTE Confidence: 0.96125424

00:14:01.635 --> 00:14:02.995 important is to add the

NOTE Confidence: 0.96125424

00:14:02.995 --> 00:14:04.835 molecular analysis for the patients

NOTE Confidence: 0.96125424

00:14:04.835 --> 00:14:06.515 because there are certain loss

NOTE Confidence: 0.96125424

00:14:06.515 --> 00:14:07.255 of heterozygosity

NOTE Confidence: 0.98757935

00:14:08.035 --> 00:14:10.435 and other chromosomal abnormalities that

NOTE Confidence: 0.98757935

00:14:10.435 --> 00:14:11.655 may determine treatment.

NOTE Confidence: 0.9428327

00:14:12.195 --> 00:14:13.155 And then you have to
NOTE Confidence: 0.9428327

00:14:13.155 --> 00:14:15.475 examine the residual kidney, the
NOTE Confidence: 0.9428327

00:14:15.475 --> 00:14:17.510 non tumor kidney. And in
NOTE Confidence: 0.9428327

00:14:17.510 --> 00:14:18.950 that, the important thing is
NOTE Confidence: 0.9428327

00:14:18.950 --> 00:14:21.110 to find nephrogenic rests because
NOTE Confidence: 0.9428327

00:14:21.110 --> 00:14:22.730 if there are nephrogenic rests,
NOTE Confidence: 0.9428327

00:14:22.790 --> 00:14:24.410 the possibility of a contralateral
NOTE Confidence: 0.8320836

00:14:25.110 --> 00:14:27.430 synchronous or metachronous once tumor
NOTE Confidence: 0.8320836

00:14:27.430 --> 00:14:28.170 is high.
NOTE Confidence: 0.97706366

00:14:28.555 --> 00:14:29.915 And these are examples that
NOTE Confidence: 0.97706366

00:14:29.915 --> 00:14:31.515 Bruce Beckwith gave me. These
NOTE Confidence: 0.97706366

00:14:31.515 --> 00:14:33.435 are pictures from him where
NOTE Confidence: 0.97706366

00:14:33.435 --> 00:14:34.655 this is the nephroblastomatosis.
NOTE Confidence: 0.9673321

00:14:35.835 --> 00:14:37.275 That word is only used
NOTE Confidence: 0.9673321

00:14:37.275 --> 00:14:38.555 for cases in which the
NOTE Confidence: 0.9673321

00:14:38.555 --> 00:14:39.855 entire renal parenchyma

NOTE Confidence: 0.92441404
00:14:40.315 --> 00:14:41.055 is nephroblastic
NOTE Confidence: 0.9695399
00:14:41.740 --> 00:14:43.260 and not a Wilms tumor
NOTE Confidence: 0.9695399
00:14:43.260 --> 00:14:44.620 directly. And this is an
NOTE Confidence: 0.9695399
00:14:44.620 --> 00:14:45.980 example of a Wilms tumor
NOTE Confidence: 0.9695399
00:14:45.980 --> 00:14:46.720 with numerous
NOTE Confidence: 0.78719074
00:14:47.100 --> 00:14:48.640 nephrogenic rest microscopically
NOTE Confidence: 0.9403483
00:14:49.180 --> 00:14:50.240 easy to identify.
NOTE Confidence: 0.9003906
00:14:50.540 --> 00:14:51.040 Okay.
NOTE Confidence: 0.9804022
00:14:51.660 --> 00:14:52.620 Let me give you an
NOTE Confidence: 0.9804022
00:14:52.620 --> 00:14:53.660 example of one of the
NOTE Confidence: 0.9804022
00:14:53.660 --> 00:14:55.020 last cases that I saw
NOTE Confidence: 0.9804022
00:14:55.020 --> 00:14:56.380 in Pittsburgh a couple of
NOTE Confidence: 0.9804022
00:14:56.380 --> 00:14:57.015 years ago.
NOTE Confidence: 0.97929454
00:14:57.655 --> 00:14:59.095 This tumor came from a
NOTE Confidence: 0.97929454
00:14:59.095 --> 00:15:00.135 two year old boy with
NOTE Confidence: 0.97929454

00:15:00.135 --> 00:15:01.255 a left renal mass. You
NOTE Confidence: 0.97929454

00:15:01.255 --> 00:15:03.115 can see the microscopic external
NOTE Confidence: 0.97929454

00:15:03.175 --> 00:15:03.675 appearance,
NOTE Confidence: 0.89038086

00:15:04.055 --> 00:15:05.675 the cross, the surface,
NOTE Confidence: 0.9515714

00:15:06.775 --> 00:15:08.295 of the, section and the
NOTE Confidence: 0.9515714

00:15:08.295 --> 00:15:10.615 residual kidney is here. Everything
NOTE Confidence: 0.9515714

00:15:10.615 --> 00:15:11.115 fine?
NOTE Confidence: 0.8479818

00:15:11.920 --> 00:15:13.060 Blastema predominance,
NOTE Confidence: 0.9586588

00:15:13.680 --> 00:15:15.940 then there is, some perirenal
NOTE Confidence: 0.9586588

00:15:16.160 --> 00:15:17.280 fat, and then there is
NOTE Confidence: 0.9586588

00:15:17.280 --> 00:15:18.580 some residual kidney.
NOTE Confidence: 0.88501793

00:15:19.280 --> 00:15:20.720 So I gave the diagnosis.
NOTE Confidence: 0.88501793

00:15:20.720 --> 00:15:21.920 It's a woman's tumor. It's
NOTE Confidence: 0.88501793

00:15:21.920 --> 00:15:23.940 a favorable histology woman's tumor,
NOTE Confidence: 0.86572266

00:15:24.315 --> 00:15:25.515 and therefore favor,
NOTE Confidence: 0.93950087

00:15:25.995 --> 00:15:27.755 the chemotherapy is not as

NOTE Confidence: 0.93950087
00:15:27.755 --> 00:15:29.675 aggressive. And the stage is
NOTE Confidence: 0.93950087
00:15:29.675 --> 00:15:30.955 stage one because it hasn't
NOTE Confidence: 0.93950087
00:15:30.955 --> 00:15:32.795 broken through the capsule or
NOTE Confidence: 0.93950087
00:15:32.795 --> 00:15:33.855 the renal signs.
NOTE Confidence: 0.93363446
00:15:36.510 --> 00:15:38.350 And then you see, you,
NOTE Confidence: 0.93363446
00:15:38.350 --> 00:15:39.550 you, you, you show the
NOTE Confidence: 0.93363446
00:15:39.550 --> 00:15:40.290 three components
NOTE Confidence: 0.94649833
00:15:41.230 --> 00:15:42.270 and then you go to
NOTE Confidence: 0.94649833
00:15:42.270 --> 00:15:42.930 the renal
NOTE Confidence: 0.7085938
00:15:43.550 --> 00:15:44.850 non tumoral parenchyma.
NOTE Confidence: 0.9988839
00:15:45.630 --> 00:15:46.910 And this is where things
NOTE Confidence: 0.9988839
00:15:46.910 --> 00:15:47.650 get interesting
NOTE Confidence: 0.5449219
00:15:48.350 --> 00:15:48.850 because
NOTE Confidence: 0.8925069
00:15:49.390 --> 00:15:50.990 diagnosing one's tumor is not
NOTE Confidence: 0.8925069
00:15:50.990 --> 00:15:51.490 difficult.
NOTE Confidence: 0.97314453

00:15:53.125 --> 00:15:54.585 But looking at this
NOTE Confidence: 0.90148926
00:15:55.285 --> 00:15:56.105 is difficult.
NOTE Confidence: 0.9887543
00:15:56.885 --> 00:15:58.245 Not looking at it, but
NOTE Confidence: 0.9887543
00:15:58.245 --> 00:15:59.845 in understanding what you are
NOTE Confidence: 0.9887543
00:15:59.845 --> 00:16:01.625 seeing. What you have here
NOTE Confidence: 0.9887543
00:16:01.685 --> 00:16:02.185 are
NOTE Confidence: 0.99786377
00:16:02.645 --> 00:16:04.425 tubular structures in the medulla
NOTE Confidence: 0.99786377
00:16:04.485 --> 00:16:05.464 that are separated
NOTE Confidence: 0.8741048
00:16:05.930 --> 00:16:07.769 by a relatively mixed soy
NOTE Confidence: 0.8741048
00:16:07.769 --> 00:16:08.269 type
NOTE Confidence: 0.7988281
00:16:08.730 --> 00:16:09.550 of stroma
NOTE Confidence: 0.99423826
00:16:10.250 --> 00:16:12.250 that is not normal. This
NOTE Confidence: 0.99423826
00:16:12.250 --> 00:16:13.870 is not the normal morphology
NOTE Confidence: 0.98673505
00:16:14.410 --> 00:16:15.870 of the renal medulla.
NOTE Confidence: 0.9936035
00:16:16.250 --> 00:16:17.709 And when you see that,
NOTE Confidence: 0.9308233
00:16:18.685 --> 00:16:20.365 you have to diagnose it.

NOTE Confidence: 0.9308233

00:16:20.365 --> 00:16:21.805 So the diagnosis that I

NOTE Confidence: 0.9308233

00:16:21.805 --> 00:16:23.245 issued in this case is

NOTE Confidence: 0.9308233

00:16:23.245 --> 00:16:25.565 re is, written here. And

NOTE Confidence: 0.9308233

00:16:25.565 --> 00:16:27.585 then I put renal medullary

NOTE Confidence: 0.9308233

00:16:27.805 --> 00:16:30.205 dysplasia consistent with Beckwith Wiedemann

NOTE Confidence: 0.9308233

00:16:30.205 --> 00:16:32.385 syndrome, see comment and references.

NOTE Confidence: 0.8313721

00:16:33.779 --> 00:16:35.300 When we discuss this case

NOTE Confidence: 0.8313721

00:16:35.300 --> 00:16:36.680 at tumor war in Pittsburgh,

NOTE Confidence: 0.8830295

00:16:38.180 --> 00:16:40.920 the, director of pediatric oncology,

NOTE Confidence: 0.8830295

00:16:41.060 --> 00:16:42.820 the clinical director of pediatric

NOTE Confidence: 0.8830295

00:16:42.820 --> 00:16:44.040 oncology there,

NOTE Confidence: 0.8877767

00:16:44.820 --> 00:16:46.180 first I said, this patient

NOTE Confidence: 0.8877767

00:16:46.180 --> 00:16:47.380 has wounds tumor and this

NOTE Confidence: 0.8877767

00:16:47.380 --> 00:16:48.500 and this and then back

NOTE Confidence: 0.8877767

00:16:48.500 --> 00:16:50.175 with William and Singh. And

NOTE Confidence: 0.8877767

00:16:50.175 --> 00:16:51.395 there was a silence.
NOTE Confidence: 0.97077286

00:16:52.575 --> 00:16:54.095 Very uncomfortable, to be honest
NOTE Confidence: 0.97077286

00:16:54.095 --> 00:16:54.755 with you.
NOTE Confidence: 0.98398435

00:16:55.375 --> 00:16:57.155 And then he said, well,
NOTE Confidence: 0.9194336

00:16:57.775 --> 00:16:58.275 maybe.
NOTE Confidence: 0.9373932

00:17:00.735 --> 00:17:02.255 But, no, it's not a
NOTE Confidence: 0.9373932

00:17:02.255 --> 00:17:03.500 maybe. It is,
NOTE Confidence: 0.8283081

00:17:04.119 --> 00:17:05.580 Beckwith Wiedemann syndrome.
NOTE Confidence: 0.98746747

00:17:05.880 --> 00:17:07.240 I put this,
NOTE Confidence: 0.9659288

00:17:07.640 --> 00:17:09.240 comment in the report, and
NOTE Confidence: 0.9659288

00:17:09.240 --> 00:17:10.460 I mentioned a previous,
NOTE Confidence: 0.9553765

00:17:11.320 --> 00:17:14.119 reference that Jorge Dotto anybody
NOTE Confidence: 0.9553765

00:17:14.119 --> 00:17:15.820 here remembers Jorge Dotto?
NOTE Confidence: 0.9675481

00:17:16.555 --> 00:17:17.595 He wrote to me this
NOTE Confidence: 0.9675481

00:17:17.595 --> 00:17:18.635 morning because he saw the
NOTE Confidence: 0.9675481

00:17:18.635 --> 00:17:19.775 announcement in Twitter,

NOTE Confidence: 0.9744014
00:17:20.315 --> 00:17:21.595 of this conference, and he
NOTE Confidence: 0.9744014
00:17:21.595 --> 00:17:23.035 said, oh, Miguel. It's nice
NOTE Confidence: 0.9744014
00:17:23.035 --> 00:17:24.155 that you are going back
NOTE Confidence: 0.9744014
00:17:24.155 --> 00:17:25.535 to Yale. Well, I'm gonna
NOTE Confidence: 0.9744014
00:17:25.675 --> 00:17:27.035 show your name because he
NOTE Confidence: 0.9744014
00:17:27.035 --> 00:17:27.695 and I
NOTE Confidence: 0.9866333
00:17:28.315 --> 00:17:29.994 published a case that,
NOTE Confidence: 0.97526044
00:17:31.440 --> 00:17:33.059 was similar to this one.
NOTE Confidence: 0.97526044
00:17:33.279 --> 00:17:34.500 The case from Pittsburgh
NOTE Confidence: 0.9980469
00:17:35.279 --> 00:17:36.179 was published
NOTE Confidence: 0.9578125
00:17:37.119 --> 00:17:38.580 with this group of authors,
NOTE Confidence: 0.93868005
00:17:38.880 --> 00:17:40.159 and one of them, Julia
NOTE Confidence: 0.93868005
00:17:40.159 --> 00:17:41.919 Mead, is a pediatric, you
NOTE Confidence: 0.93868005
00:17:41.919 --> 00:17:43.539 know, Julia Mead, Rosanna.
NOTE Confidence: 0.9676723
00:17:43.905 --> 00:17:45.585 She's a pediatric oncologist that
NOTE Confidence: 0.9676723

00:17:45.585 --> 00:17:47.445 is specialized in cancer predisposition

NOTE Confidence: 0.9676723

00:17:47.665 --> 00:17:49.665 syndromes. And, the following day

NOTE Confidence: 0.9676723

00:17:49.665 --> 00:17:50.465 of the tumor board, she

NOTE Confidence: 0.9676723

00:17:50.465 --> 00:17:51.425 comes to my office and

NOTE Confidence: 0.9676723

00:17:51.425 --> 00:17:53.025 she says, Miguel, can I

NOTE Confidence: 0.9676723

00:17:53.025 --> 00:17:54.005 cry with you?

NOTE Confidence: 0.9975586

00:17:54.545 --> 00:17:55.285 What happened?

NOTE Confidence: 0.93892694

00:17:56.320 --> 00:17:57.680 They you were right. They

NOTE Confidence: 0.93892694

00:17:57.680 --> 00:17:59.600 missed it. And the problem

NOTE Confidence: 0.93892694

00:17:59.600 --> 00:18:01.200 is that with, Beckwith Wiedemann

NOTE Confidence: 0.93892694

00:18:01.200 --> 00:18:02.400 syndrome is not an easy

NOTE Confidence: 0.93892694

00:18:02.400 --> 00:18:04.740 diagnosis to make even clinically,

NOTE Confidence: 0.93892694

00:18:04.880 --> 00:18:06.100 even to the specialist.

NOTE Confidence: 0.975293

00:18:06.800 --> 00:18:08.820 I remember an autopsy here

NOTE Confidence: 0.97233075

00:18:10.054 --> 00:18:11.434 that one of our geneticists,

NOTE Confidence: 0.92770386

00:18:11.734 --> 00:18:14.155 a very highly respected geneticists

NOTE Confidence: 0.92770386
00:18:14.375 --> 00:18:16.135 here came to see. And
NOTE Confidence: 0.92770386
00:18:16.135 --> 00:18:16.794 when I
NOTE Confidence: 0.88725936
00:18:17.095 --> 00:18:18.135 just looked at it and
NOTE Confidence: 0.88725936
00:18:18.135 --> 00:18:18.855 I said, oh, it's a
NOTE Confidence: 0.88725936
00:18:18.855 --> 00:18:20.234 Beckwith William and syndrome.
NOTE Confidence: 0.98136395
00:18:20.535 --> 00:18:21.655 She looked at me like,
NOTE Confidence: 0.98136395
00:18:21.655 --> 00:18:22.155 what?
NOTE Confidence: 0.95289713
00:18:22.519 --> 00:18:24.119 And it was because it's
NOTE Confidence: 0.95289713
00:18:24.119 --> 00:18:25.399 not easy to diagnose it
NOTE Confidence: 0.95289713
00:18:25.399 --> 00:18:26.940 unless you think of it.
NOTE Confidence: 0.9921875
00:18:27.720 --> 00:18:28.220 So
NOTE Confidence: 0.95666504
00:18:29.080 --> 00:18:30.600 this patient that I show
NOTE Confidence: 0.95666504
00:18:30.600 --> 00:18:31.659 you was published
NOTE Confidence: 0.9364746
00:18:32.039 --> 00:18:34.299 and showing the medullary race,
NOTE Confidence: 0.9945746
00:18:34.679 --> 00:18:36.299 the medullary dysplasia,
NOTE Confidence: 0.95016277

00:18:36.759 --> 00:18:39.045 all the correlation with radiology,
NOTE Confidence: 0.93632

00:18:39.345 --> 00:18:41.345 and then other examples in
NOTE Confidence: 0.93632

00:18:41.345 --> 00:18:41.845 our,
NOTE Confidence: 0.73583984

00:18:43.425 --> 00:18:43.925 files
NOTE Confidence: 0.9258968

00:18:44.225 --> 00:18:45.905 were found with Beckwith Guillian
NOTE Confidence: 0.9258968

00:18:45.905 --> 00:18:47.744 Syndrome and Guillian's tumor. What
NOTE Confidence: 0.9258968

00:18:47.744 --> 00:18:49.345 is Beckwith Guillian Syndrome? Well,
NOTE Confidence: 0.9258968

00:18:49.345 --> 00:18:51.080 you know, it's a combination
NOTE Confidence: 0.9258968

00:18:51.140 --> 00:18:51.799 of overgrowth
NOTE Confidence: 0.9051819

00:18:52.260 --> 00:18:54.820 affecting multiple organs described by
NOTE Confidence: 0.9051819

00:18:54.820 --> 00:18:56.200 Bruce Beckwith and,
NOTE Confidence: 0.9188477

00:18:56.500 --> 00:18:57.400 Hans Wiedemann.
NOTE Confidence: 0.9195731

00:18:58.179 --> 00:18:59.220 On a case report, by
NOTE Confidence: 0.9195731

00:18:59.220 --> 00:19:00.179 the way, and there are
NOTE Confidence: 0.9195731

00:19:00.179 --> 00:19:01.720 people that are very,
NOTE Confidence: 0.93036246

00:19:02.340 --> 00:19:03.460 upset when you say I'm

NOTE Confidence: 0.93036246
00:19:03.460 --> 00:19:04.340 going to do a case
NOTE Confidence: 0.93036246
00:19:04.340 --> 00:19:05.835 report and say what? You
NOTE Confidence: 0.93036246
00:19:05.835 --> 00:19:06.075 should that
NOTE Confidence: 0.9991455
00:19:06.955 --> 00:19:08.335 that's that's not good.
NOTE Confidence: 0.99690753
00:19:08.875 --> 00:19:09.775 Well, if Bruce
NOTE Confidence: 0.86469185
00:19:10.155 --> 00:19:11.674 of Witten or Witherman did
NOTE Confidence: 0.86469185
00:19:11.674 --> 00:19:13.054 not report that case,
NOTE Confidence: 0.97698975
00:19:13.674 --> 00:19:16.174 the field of overgrowth syndromes
NOTE Confidence: 0.97698975
00:19:16.315 --> 00:19:18.095 would not have started. Okay?
NOTE Confidence: 0.97698975
00:19:18.230 --> 00:19:18.890 So, yes,
NOTE Confidence: 0.91532135
00:19:19.270 --> 00:19:20.550 you may be wasting your
NOTE Confidence: 0.91532135
00:19:20.550 --> 00:19:22.230 time as I spoke this
NOTE Confidence: 0.91532135
00:19:22.230 --> 00:19:23.990 morning with doctor Ho how,
NOTE Confidence: 0.91532135
00:19:24.310 --> 00:19:24.810 go,
NOTE Confidence: 0.98783505
00:19:25.270 --> 00:19:26.950 on case reports, but you
NOTE Confidence: 0.98783505

00:19:26.950 --> 00:19:27.990 may not if you ask
NOTE Confidence: 0.98783505

00:19:27.990 --> 00:19:30.070 the interesting question. So don't
NOTE Confidence: 0.98783505

00:19:30.070 --> 00:19:31.910 don't don't get dissuaded to
NOTE Confidence: 0.98783505

00:19:31.910 --> 00:19:33.505 write cases if you think
NOTE Confidence: 0.98783505

00:19:33.505 --> 00:19:34.565 that they are interesting.
NOTE Confidence: 0.961084

00:19:34.945 --> 00:19:36.165 These are all the genetic
NOTE Confidence: 0.961084

00:19:36.305 --> 00:19:37.905 abnormalities. I'm not going to
NOTE Confidence: 0.961084

00:19:37.905 --> 00:19:39.345 detail them because this is
NOTE Confidence: 0.961084

00:19:39.345 --> 00:19:41.185 not the function, of this,
NOTE Confidence: 0.961084

00:19:41.585 --> 00:19:42.705 conference. It's just to call
NOTE Confidence: 0.961084

00:19:42.705 --> 00:19:44.065 your attention at what you
NOTE Confidence: 0.961084

00:19:44.065 --> 00:19:45.585 need to look under the
NOTE Confidence: 0.961084

00:19:45.585 --> 00:19:47.205 microscope or at the microscopic
NOTE Confidence: 1

00:19:47.505 --> 00:19:48.005 level.
NOTE Confidence: 0.8264567

00:19:49.640 --> 00:19:51.240 This is an example of
NOTE Confidence: 0.8264567

00:19:51.240 --> 00:19:52.600 a fetus with with a

NOTE Confidence: 0.8264567
00:19:52.600 --> 00:19:53.340 man syndrome
NOTE Confidence: 0.9723145
00:19:54.359 --> 00:19:56.119 that we, carried on an
NOTE Confidence: 0.9723145
00:19:56.119 --> 00:19:58.540 autopsy here many years ago.
NOTE Confidence: 0.9739014
00:19:58.920 --> 00:20:00.059 You can see the exome
NOTE Confidence: 0.9739014
00:20:00.119 --> 00:20:01.660 follows. This is not umbilical
NOTE Confidence: 0.98531896
00:20:02.244 --> 00:20:03.684 hernia, but it's a huge
NOTE Confidence: 0.98531896
00:20:03.684 --> 00:20:06.424 umbilical hernia hernia that contains
NOTE Confidence: 0.95162684
00:20:06.725 --> 00:20:08.645 all the abdominal organs, including
NOTE Confidence: 0.95162684
00:20:08.645 --> 00:20:09.924 the liver. And you see
NOTE Confidence: 0.95162684
00:20:09.924 --> 00:20:11.465 the the the two fetuses
NOTE Confidence: 0.95162684
00:20:11.524 --> 00:20:13.765 here with coarse features. They
NOTE Confidence: 0.95162684
00:20:13.765 --> 00:20:14.490 don't look
NOTE Confidence: 0.9996745
00:20:14.890 --> 00:20:16.330 nice. They are not cute
NOTE Confidence: 0.9996745
00:20:16.330 --> 00:20:16.830 babies.
NOTE Confidence: 0.9830322
00:20:17.290 --> 00:20:18.830 They are coarse because
NOTE Confidence: 0.99853516

00:20:19.369 --> 00:20:20.109 the overgrowth
NOTE Confidence: 0.94240993

00:20:20.490 --> 00:20:22.090 of the tissues, the soft
NOTE Confidence: 0.94240993

00:20:22.090 --> 00:20:23.950 tissues produces these creases
NOTE Confidence: 0.99658203

00:20:24.490 --> 00:20:25.230 and foldings
NOTE Confidence: 0.9984375

00:20:25.690 --> 00:20:27.550 that make them look not
NOTE Confidence: 0.9980469

00:20:28.225 --> 00:20:28.965 not cute.
NOTE Confidence: 0.9275462

00:20:29.505 --> 00:20:30.784 They have these creases in
NOTE Confidence: 0.9275462

00:20:30.784 --> 00:20:31.904 the ears as you can
NOTE Confidence: 0.9275462

00:20:31.904 --> 00:20:33.664 see here, these fissures and
NOTE Confidence: 0.9275462

00:20:33.664 --> 00:20:34.164 creases,
NOTE Confidence: 0.96612257

00:20:34.465 --> 00:20:36.065 and they obviously have different
NOTE Confidence: 0.96612257

00:20:36.065 --> 00:20:37.825 degrees of umbilical cord. And
NOTE Confidence: 0.96612257

00:20:37.825 --> 00:20:39.205 they can have hemihypertrophy
NOTE Confidence: 0.99674475

00:20:40.065 --> 00:20:40.725 or hypertrophy
NOTE Confidence: 0.99560547

00:20:41.105 --> 00:20:41.845 of something.
NOTE Confidence: 0.938337

00:20:42.540 --> 00:20:44.060 When I invited Bruce Beckwith

NOTE Confidence: 0.938337
00:20:44.060 --> 00:20:45.500 to give Grand Rounds right
NOTE Confidence: 0.938337
00:20:45.500 --> 00:20:47.119 here many years ago,
NOTE Confidence: 0.9633179
00:20:47.580 --> 00:20:49.740 we had lunch together, and
NOTE Confidence: 0.9633179
00:20:49.740 --> 00:20:50.940 he told me, you know,
NOTE Confidence: 0.9633179
00:20:50.940 --> 00:20:51.600 the most,
NOTE Confidence: 0.9941406
00:20:53.740 --> 00:20:54.240 unforgettable
NOTE Confidence: 0.89228314
00:20:54.619 --> 00:20:56.255 case of she never said
NOTE Confidence: 0.89228314
00:20:56.255 --> 00:20:58.095 Beckwith Wiedemann syndrome. He said
NOTE Confidence: 0.89228314
00:20:58.095 --> 00:20:58.914 of BWS
NOTE Confidence: 0.99263823
00:21:00.255 --> 00:21:01.615 was a case that I
NOTE Confidence: 0.99263823
00:21:01.615 --> 00:21:02.734 saw on a man with
NOTE Confidence: 0.99263823
00:21:02.734 --> 00:21:03.794 a huge thumb.
NOTE Confidence: 0.88516355
00:21:04.654 --> 00:21:06.734 Okay? Because he doesn't hit
NOTE Confidence: 0.88516355
00:21:06.734 --> 00:21:08.414 the the normality, doesn't hit
NOTE Confidence: 0.88516355
00:21:08.414 --> 00:21:10.015 all the organs at the
NOTE Confidence: 0.88516355

00:21:10.015 --> 00:21:11.270 same level. Okay?

NOTE Confidence: 0.8828125

00:21:12.290 --> 00:21:12.790 The

NOTE Confidence: 0.99201083

00:21:13.810 --> 00:21:15.990 fetal cortex in the adrenal

NOTE Confidence: 0.99201083

00:21:16.050 --> 00:21:18.070 medulla looks like this normally.

NOTE Confidence: 0.99201083

00:21:18.369 --> 00:21:19.970 This is the permanent cortex

NOTE Confidence: 0.99201083

00:21:19.970 --> 00:21:21.350 in the process of development.

NOTE Confidence: 0.99201083

00:21:21.410 --> 00:21:22.530 This is a fetus or

NOTE Confidence: 0.99201083

00:21:22.530 --> 00:21:24.130 a baby of less than

NOTE Confidence: 0.99201083

00:21:24.130 --> 00:21:25.910 six months of age postnatal.

NOTE Confidence: 0.9599365

00:21:26.575 --> 00:21:27.775 Okay? But it can be

NOTE Confidence: 0.9599365

00:21:27.775 --> 00:21:29.795 a newborn. And the pancreas

NOTE Confidence: 0.9206543

00:21:30.255 --> 00:21:31.775 may look like this. In

NOTE Confidence: 0.9206543

00:21:31.775 --> 00:21:33.615 Beckwith Wiedemann syndrome cases, you

NOTE Confidence: 0.9206543

00:21:33.615 --> 00:21:34.115 see

NOTE Confidence: 0.9654541

00:21:34.895 --> 00:21:35.395 nucleomegaly

NOTE Confidence: 0.9934082

00:21:35.935 --> 00:21:37.795 in the fetal adrenal cortex.

NOTE Confidence: 0.99902344
00:21:38.255 --> 00:21:40.095 That is a sign that
NOTE Confidence: 0.99902344
00:21:40.095 --> 00:21:41.635 is frequently associated
NOTE Confidence: 0.95701414
00:21:42.289 --> 00:21:44.770 to Beckwith Wiedemann syndrome. Not
NOTE Confidence: 0.95701414
00:21:44.770 --> 00:21:46.770 always. Sometimes it's just a
NOTE Confidence: 0.95701414
00:21:46.770 --> 00:21:48.049 sign of stress, but when
NOTE Confidence: 0.95701414
00:21:48.049 --> 00:21:49.090 it is when it is
NOTE Confidence: 0.95701414
00:21:49.090 --> 00:21:51.250 so diffuse, that's another clue
NOTE Confidence: 0.95701414
00:21:51.250 --> 00:21:52.869 that you should not miss.
NOTE Confidence: 0.95701414
00:21:53.010 --> 00:21:54.049 Because even if you are
NOTE Confidence: 0.95701414
00:21:54.049 --> 00:21:55.409 doing an autopsy or if
NOTE Confidence: 0.95701414
00:21:55.409 --> 00:21:57.075 you get a nephrectomy with
NOTE Confidence: 0.95701414
00:21:57.154 --> 00:21:58.434 an adrenal gland that looks
NOTE Confidence: 0.95701414
00:21:58.434 --> 00:21:59.635 like that, you need to
NOTE Confidence: 0.95701414
00:21:59.635 --> 00:22:00.774 call doctor
NOTE Confidence: 0.80159503
00:22:01.794 --> 00:22:02.294 Farzana
NOTE Confidence: 0.8684082

00:22:02.595 --> 00:22:03.255 and say,
NOTE Confidence: 0.8759591

00:22:04.514 --> 00:22:06.674 Beckwith Wiedemann syndrome. Please check
NOTE Confidence: 0.8759591

00:22:06.674 --> 00:22:07.414 for that.
NOTE Confidence: 0.94454974

00:22:09.790 --> 00:22:11.470 The CD of blastosis is
NOTE Confidence: 0.94454974

00:22:11.470 --> 00:22:12.830 the process of formation of
NOTE Confidence: 0.94454974

00:22:12.830 --> 00:22:14.510 the islets of Langerhans. And
NOTE Confidence: 0.94454974

00:22:14.510 --> 00:22:16.270 in Beckwith Wiedemann syndrome, you've
NOTE Confidence: 0.94454974

00:22:16.270 --> 00:22:17.150 had a you have an
NOTE Confidence: 0.94454974

00:22:17.150 --> 00:22:19.390 increased number of islets of
NOTE Confidence: 0.94454974

00:22:19.390 --> 00:22:21.090 Langerhans. This is the case
NOTE Confidence: 0.94454974

00:22:21.230 --> 00:22:22.990 that I published with, Jorge
NOTE Confidence: 0.94454974

00:22:22.990 --> 00:22:23.950 Dotto. And let me tell
NOTE Confidence: 0.94454974

00:22:23.950 --> 00:22:25.184 you the story. When when
NOTE Confidence: 0.94454974

00:22:25.184 --> 00:22:26.465 this you were not part
NOTE Confidence: 0.94454974

00:22:26.465 --> 00:22:27.505 of the clinical team at
NOTE Confidence: 0.94454974

00:22:27.505 --> 00:22:28.885 that time, I think, but

NOTE Confidence: 0.94454974
00:22:28.945 --> 00:22:30.784 they came to the search
NOTE Confidence: 0.94454974
00:22:30.784 --> 00:22:32.225 pad area, and I was
NOTE Confidence: 0.94454974
00:22:32.225 --> 00:22:33.585 sitting with Jorge looking at
NOTE Confidence: 0.94454974
00:22:33.585 --> 00:22:34.244 the case.
NOTE Confidence: 0.94142103
00:22:34.705 --> 00:22:36.304 And when, they asked me,
NOTE Confidence: 0.94142103
00:22:36.304 --> 00:22:37.105 what is it? Is it
NOTE Confidence: 0.94142103
00:22:37.105 --> 00:22:37.984 the Wilms? And I said,
NOTE Confidence: 0.94142103
00:22:37.984 --> 00:22:38.910 yes. It is a one's
NOTE Confidence: 0.94142103
00:22:38.990 --> 00:22:40.109 tumor. Is it favorable? Yes.
NOTE Confidence: 0.94142103
00:22:40.109 --> 00:22:41.150 It is favorable. Thank you
NOTE Confidence: 0.94142103
00:22:41.150 --> 00:22:42.210 very much. Okay.
NOTE Confidence: 0.99296874
00:22:42.590 --> 00:22:44.130 And then when I continue
NOTE Confidence: 0.9963379
00:22:44.510 --> 00:22:45.970 looking at the normal
NOTE Confidence: 0.9647624
00:22:46.350 --> 00:22:48.510 residual kidney, it is not
NOTE Confidence: 0.9647624
00:22:48.510 --> 00:22:49.010 normal.
NOTE Confidence: 0.9370972

00:22:49.550 --> 00:22:51.310 And I I asked Jorge
NOTE Confidence: 0.9370972

00:22:51.310 --> 00:22:52.670 and other residents that work
NOTE Confidence: 0.9370972

00:22:52.670 --> 00:22:54.190 with me to walk to
NOTE Confidence: 0.9370972

00:22:54.190 --> 00:22:55.725 the bed of the patient.
NOTE Confidence: 0.9598675

00:22:56.825 --> 00:22:58.904 Our our clinical colleagues looked
NOTE Confidence: 0.9598675

00:22:58.904 --> 00:22:59.945 at us like, what are
NOTE Confidence: 0.9598675

00:22:59.945 --> 00:23:01.065 these guys doing here on
NOTE Confidence: 0.9598675

00:23:01.065 --> 00:23:02.524 the floor? These are pathologists.
NOTE Confidence: 0.9598675

00:23:02.744 --> 00:23:04.265 Right? Well, I I asked
NOTE Confidence: 0.9598675

00:23:04.265 --> 00:23:05.244 permission to
NOTE Confidence: 0.9816081

00:23:05.945 --> 00:23:07.164 check the external,
NOTE Confidence: 0.8918457

00:23:08.650 --> 00:23:09.390 the the clinical,
NOTE Confidence: 0.95186675

00:23:10.010 --> 00:23:11.130 appearance of the of the
NOTE Confidence: 0.95186675

00:23:11.130 --> 00:23:12.730 of the patient. He was
NOTE Confidence: 0.95186675

00:23:12.730 --> 00:23:13.950 a cute baby,
NOTE Confidence: 0.92510986

00:23:15.050 --> 00:23:16.350 about a year old,

NOTE Confidence: 0.9338379
00:23:17.210 --> 00:23:17.710 and,
NOTE Confidence: 0.86185825
00:23:18.970 --> 00:23:20.505 didn't have anything that looked
NOTE Confidence: 0.86185825
00:23:20.585 --> 00:23:22.685 like Benjamin syndrome. No hemihypertrophy,
NOTE Confidence: 0.8998284
00:23:23.385 --> 00:23:24.984 no creases, nothing, not this
NOTE Confidence: 0.8998284
00:23:24.984 --> 00:23:26.425 morphology. And I was very
NOTE Confidence: 0.8998284
00:23:26.425 --> 00:23:26.925 disappointed.
NOTE Confidence: 0.93725586
00:23:27.705 --> 00:23:28.525 And I said,
NOTE Confidence: 0.98115236
00:23:29.145 --> 00:23:30.345 can't be. I sent a
NOTE Confidence: 0.98115236
00:23:30.345 --> 00:23:31.885 couple of pictures to Bruce
NOTE Confidence: 0.9363237
00:23:32.185 --> 00:23:33.225 and he calls me and
NOTE Confidence: 0.9363237
00:23:33.225 --> 00:23:35.429 he says, your patient has
NOTE Confidence: 0.9363237
00:23:35.429 --> 00:23:35.929 PWS
NOTE Confidence: 0.7216797
00:23:36.230 --> 00:23:36.730 whether
NOTE Confidence: 0.97751194
00:23:37.109 --> 00:23:38.309 the rest of his body
NOTE Confidence: 0.97751194
00:23:38.309 --> 00:23:39.609 knows it or not.
NOTE Confidence: 0.9923828

00:23:40.470 --> 00:23:42.650 And they prove it genetically.
NOTE Confidence: 0.99902344

00:23:43.590 --> 00:23:44.090 So
NOTE Confidence: 0.9581822

00:23:44.550 --> 00:23:45.770 this is a type of
NOTE Confidence: 0.9581822

00:23:45.830 --> 00:23:47.050 malformation or
NOTE Confidence: 0.9970703

00:23:47.655 --> 00:23:48.155 abnormal
NOTE Confidence: 0.9193344

00:23:48.615 --> 00:23:50.295 development of the renal medulla
NOTE Confidence: 0.9193344

00:23:50.295 --> 00:23:51.494 that you can appreciate in
NOTE Confidence: 0.9193344

00:23:51.494 --> 00:23:53.175 Beckwith Wiedemann syndrome with this
NOTE Confidence: 0.9193344

00:23:53.175 --> 00:23:53.675 mixoid
NOTE Confidence: 0.9688069

00:23:54.215 --> 00:23:55.815 stroma around the tubules and
NOTE Confidence: 0.9688069

00:23:55.815 --> 00:23:57.515 the separation of the tubules.
NOTE Confidence: 0.95606446

00:23:57.895 --> 00:23:59.430 Please pay attention to that
NOTE Confidence: 0.95606446

00:23:59.510 --> 00:24:00.710 in those cases. You can
NOTE Confidence: 0.95606446

00:24:00.710 --> 00:24:02.310 also see nephrogenic rest. This
NOTE Confidence: 0.95606446

00:24:02.310 --> 00:24:03.910 is another example with a
NOTE Confidence: 0.95606446

00:24:03.910 --> 00:24:05.750 medullary ray, and these are

NOTE Confidence: 0.95606446

00:24:05.750 --> 00:24:07.850 the usual molecular subtypes

NOTE Confidence: 0.88215774

00:24:08.310 --> 00:24:09.990 of Beckwith Wiedemann syndrome. This

NOTE Confidence: 0.88215774

00:24:09.990 --> 00:24:11.830 is taken from the pits

NOTE Confidence: 0.88215774

00:24:11.830 --> 00:24:12.230 five,

NOTE Confidence: 0.7831421

00:24:13.030 --> 00:24:14.330 edition of the WHO,

NOTE Confidence: 0.9847819

00:24:15.805 --> 00:24:17.105 classification of tumors,

NOTE Confidence: 0.9691337

00:24:17.565 --> 00:24:19.165 and we put these pictures

NOTE Confidence: 0.9691337

00:24:19.165 --> 00:24:19.825 in that,

NOTE Confidence: 1

00:24:20.365 --> 00:24:20.865 volume.

NOTE Confidence: 0.9359131

00:24:21.645 --> 00:24:23.265 Other tumors may also,

NOTE Confidence: 0.93358505

00:24:24.685 --> 00:24:25.885 appear in patients with with

NOTE Confidence: 0.93358505

00:24:25.885 --> 00:24:27.809 Beckwith Wiedemann syndrome, of course.

NOTE Confidence: 0.93358505

00:24:27.809 --> 00:24:28.789 This is a case,

NOTE Confidence: 0.9442302

00:24:29.490 --> 00:24:32.630 that, had a, mesenchymal hamartoma

NOTE Confidence: 0.9764404

00:24:33.250 --> 00:24:34.130 associated with,

NOTE Confidence: 0.9314733

00:24:34.609 --> 00:24:36.130 Beckwith Wiedemann syndrome. By the
NOTE Confidence: 0.9314733

00:24:36.130 --> 00:24:37.590 way, the alpha fetoprotein
NOTE Confidence: 0.9613444

00:24:37.890 --> 00:24:38.929 in this patient was a
NOTE Confidence: 0.9613444

00:24:38.929 --> 00:24:39.429 million,
NOTE Confidence: 0.9144694

00:24:39.895 --> 00:24:40.955 above a million.
NOTE Confidence: 0.96432287

00:24:41.575 --> 00:24:42.615 And it was not a
NOTE Confidence: 0.96432287

00:24:42.615 --> 00:24:43.115 hepatoblastoma,
NOTE Confidence: 0.99902344

00:24:43.575 --> 00:24:44.075 which
NOTE Confidence: 0.8859863

00:24:44.375 --> 00:24:45.575 what would have been the
NOTE Confidence: 0.8859863

00:24:45.575 --> 00:24:46.635 first suspicion.
NOTE Confidence: 0.9880452

00:24:47.415 --> 00:24:48.695 Okay? But it's not a
NOTE Confidence: 0.9880452

00:24:48.695 --> 00:24:49.195 hepatoblastoma.
NOTE Confidence: 0.9719238

00:24:49.494 --> 00:24:50.535 I remember we sent it
NOTE Confidence: 0.9719238

00:24:50.535 --> 00:24:51.755 to Milton Feingold
NOTE Confidence: 0.9058431

00:24:52.250 --> 00:24:54.170 at Texas Children's Hospital, and
NOTE Confidence: 0.9058431

00:24:54.170 --> 00:24:55.050 he called me. He said,

NOTE Confidence: 0.9058431
00:24:55.050 --> 00:24:56.510 you're right, Miguel. He said,
NOTE Confidence: 0.9733887
00:24:58.210 --> 00:24:59.050 So we need to pay
NOTE Confidence: 0.9733887
00:24:59.050 --> 00:25:00.810 attention because the morphology of
NOTE Confidence: 0.9733887
00:25:00.810 --> 00:25:01.550 these lesions
NOTE Confidence: 0.9970703
00:25:02.010 --> 00:25:02.510 may
NOTE Confidence: 0.9980469
00:25:03.130 --> 00:25:04.490 give you the clue and
NOTE Confidence: 0.9980469
00:25:04.490 --> 00:25:05.150 the diagnosis
NOTE Confidence: 0.94940186
00:25:06.215 --> 00:25:08.135 of Beckwith Wiedemann syndrome. Let's
NOTE Confidence: 0.94940186
00:25:08.135 --> 00:25:08.635 change
NOTE Confidence: 0.96859974
00:25:11.494 --> 00:25:13.015 organs. I'm sure that you
NOTE Confidence: 0.96859974
00:25:13.015 --> 00:25:15.175 all have heard, used, and
NOTE Confidence: 0.96859974
00:25:15.175 --> 00:25:16.395 applied the term
NOTE Confidence: 0.9333496
00:25:16.775 --> 00:25:17.275 CPAM
NOTE Confidence: 0.97444665
00:25:17.815 --> 00:25:18.715 or CCAM.
NOTE Confidence: 0.9801432
00:25:19.340 --> 00:25:20.560 Right? Cystic,
NOTE Confidence: 0.9180664

00:25:22.380 --> 00:25:22.880 adenomatoid
NOTE Confidence: 0.9281529

00:25:23.260 --> 00:25:25.100 malformation of the lung or
NOTE Confidence: 0.9281529

00:25:25.100 --> 00:25:26.000 cystic pulmonary,
NOTE Confidence: 0.99902344

00:25:27.100 --> 00:25:27.600 malformation
NOTE Confidence: 0.99853516

00:25:27.900 --> 00:25:28.640 of the lung.
NOTE Confidence: 0.99853516

00:25:29.100 --> 00:25:29.600 Well,
NOTE Confidence: 0.99677736

00:25:30.780 --> 00:25:32.240 please don't use it anymore.
NOTE Confidence: 1

00:25:33.465 --> 00:25:33.965 It's
NOTE Confidence: 0.9681315

00:25:34.265 --> 00:25:36.345 changing. The nomenclature is being
NOTE Confidence: 0.9681315

00:25:36.345 --> 00:25:37.865 updated. I haven't made that
NOTE Confidence: 0.9681315

00:25:37.865 --> 00:25:39.805 diagnosis in about fourteen years.
NOTE Confidence: 0.9681315

00:25:40.105 --> 00:25:41.565 There are people that write
NOTE Confidence: 0.9681315

00:25:41.785 --> 00:25:43.244 and that are very nice
NOTE Confidence: 0.9681315

00:25:43.465 --> 00:25:45.225 and trying to please people
NOTE Confidence: 0.9681315

00:25:45.225 --> 00:25:47.145 that have described this. Among
NOTE Confidence: 0.9681315

00:25:47.145 --> 00:25:49.019 them, a good friend, Doctor.

NOTE Confidence: 0.9681315
00:25:49.019 --> 00:25:51.340 Tom Stalker, a retired professor
NOTE Confidence: 0.9681315
00:25:51.340 --> 00:25:53.679 who made all these very
NOTE Confidence: 0.9033203
00:25:54.299 --> 00:25:54.799 intuitive,
NOTE Confidence: 0.7783203
00:25:55.740 --> 00:25:56.240 beautiful,
NOTE Confidence: 0.99902344
00:25:56.940 --> 00:25:57.440 conceptual
NOTE Confidence: 0.9838867
00:25:58.139 --> 00:25:58.639 understanding
NOTE Confidence: 0.9525655
00:25:59.179 --> 00:26:00.925 of the development of the
NOTE Confidence: 0.9525655
00:26:00.925 --> 00:26:02.365 long and the cystic lesions.
NOTE Confidence: 0.9525655
00:26:02.365 --> 00:26:03.805 The problem is that however
NOTE Confidence: 0.9525655
00:26:03.805 --> 00:26:06.125 beautiful this was in types
NOTE Confidence: 0.9525655
00:26:06.125 --> 00:26:07.984 zero to type four,
NOTE Confidence: 0.95410156
00:26:08.605 --> 00:26:09.585 it's wrong.
NOTE Confidence: 0.97074383
00:26:10.845 --> 00:26:12.525 And, therefore, we need to
NOTE Confidence: 0.97074383
00:26:12.525 --> 00:26:13.025 move
NOTE Confidence: 0.9995931
00:26:13.490 --> 00:26:15.170 to use a more current
NOTE Confidence: 0.9995931

00:26:15.170 --> 00:26:15.670 classification.
NOTE Confidence: 0.9016182

00:26:16.609 --> 00:26:18.790 There are classically three types,
NOTE Confidence: 0.9016182

00:26:19.010 --> 00:26:21.010 large c's, medium sized c's,
NOTE Confidence: 0.9016182

00:26:21.010 --> 00:26:22.609 small c's. And then he
NOTE Confidence: 0.9016182

00:26:22.609 --> 00:26:24.290 said, well, this occur because
NOTE Confidence: 0.9016182

00:26:24.290 --> 00:26:25.725 they develop in the
NOTE Confidence: 0.85465497

00:26:26.205 --> 00:26:26.705 bronchi,
NOTE Confidence: 0.95040685

00:26:27.085 --> 00:26:29.484 terminal bronchioles, or alveoli. Well,
NOTE Confidence: 0.95040685

00:26:29.484 --> 00:26:29.804 no.
NOTE Confidence: 0.9114787

00:26:30.445 --> 00:26:31.804 She added two more types,
NOTE Confidence: 0.9114787

00:26:31.804 --> 00:26:32.304 etcetera.
NOTE Confidence: 0.9995117

00:26:32.605 --> 00:26:33.105 No.
NOTE Confidence: 0.9512952

00:26:33.885 --> 00:26:35.244 There are other things that
NOTE Confidence: 0.9512952

00:26:35.244 --> 00:26:37.005 look a little bit different
NOTE Confidence: 0.9512952

00:26:37.005 --> 00:26:37.804 but are part of the
NOTE Confidence: 0.9512952

00:26:37.804 --> 00:26:40.065 same spectrum. Congenital lower emphysema

NOTE Confidence: 0.9512952
00:26:40.125 --> 00:26:41.669 is one of them. This
NOTE Confidence: 0.9512952
00:26:41.669 --> 00:26:43.909 dilatation of the lung with
NOTE Confidence: 0.9512952
00:26:43.909 --> 00:26:44.490 a hyper,
NOTE Confidence: 0.8178711
00:26:47.029 --> 00:26:47.529 distension
NOTE Confidence: 0.94283557
00:26:48.070 --> 00:26:49.109 and you can see the
NOTE Confidence: 0.94283557
00:26:49.109 --> 00:26:50.869 microscopic appearance of this long
NOTE Confidence: 0.94283557
00:26:50.869 --> 00:26:51.369 parenchyma.
NOTE Confidence: 0.95147455
00:26:51.909 --> 00:26:53.590 And sometimes you can see
NOTE Confidence: 0.95147455
00:26:53.590 --> 00:26:55.450 like this under the diaphragm.
NOTE Confidence: 0.94929504
00:26:56.414 --> 00:26:57.215 There is a piece of
NOTE Confidence: 0.94929504
00:26:57.215 --> 00:26:58.414 lung that is very solid.
NOTE Confidence: 0.94929504
00:26:58.414 --> 00:26:59.534 This is called the Rocky
NOTE Confidence: 0.94929504
00:26:59.534 --> 00:27:00.034 Tanski
NOTE Confidence: 0.9482422
00:27:00.494 --> 00:27:00.994 lobe
NOTE Confidence: 0.96698
00:27:01.455 --> 00:27:02.894 of the lung, which is
NOTE Confidence: 0.96698

00:27:02.894 --> 00:27:04.014 part of the,
NOTE Confidence: 0.99902344

00:27:04.654 --> 00:27:05.875 spectrum of disorders
NOTE Confidence: 0.9851074

00:27:06.414 --> 00:27:07.154 that includes
NOTE Confidence: 0.94140625

00:27:07.615 --> 00:27:08.115 sequestrations,
NOTE Confidence: 0.935376

00:27:09.510 --> 00:27:11.109 so called or formerly called
NOTE Confidence: 0.935376

00:27:11.109 --> 00:27:13.369 CPAMs, and congenital lower emphysema.
NOTE Confidence: 0.935376

00:27:13.510 --> 00:27:14.790 All of those lesions come
NOTE Confidence: 0.935376

00:27:14.790 --> 00:27:16.330 to us pediatric pathologies
NOTE Confidence: 0.952492

00:27:16.869 --> 00:27:18.790 very frequently. I don't know,
NOTE Confidence: 0.952492

00:27:18.790 --> 00:27:20.250 Rafaela. You must receive
NOTE Confidence: 0.9992676

00:27:20.550 --> 00:27:22.010 about one every month
NOTE Confidence: 0.96580595

00:27:22.755 --> 00:27:25.234 probably or more. Right? Because
NOTE Confidence: 0.96580595

00:27:25.234 --> 00:27:26.275 that's more or less what
NOTE Confidence: 0.96580595

00:27:26.275 --> 00:27:27.395 I remember and they look
NOTE Confidence: 0.96580595

00:27:27.395 --> 00:27:28.055 like this.
NOTE Confidence: 0.9980469

00:27:28.675 --> 00:27:29.494 And we

NOTE Confidence: 0.997454
00:27:29.795 --> 00:27:31.234 usually don't have a problem
NOTE Confidence: 0.997454
00:27:31.234 --> 00:27:32.455 diagnosing this.
NOTE Confidence: 0.9995117
00:27:32.915 --> 00:27:33.655 The problem
NOTE Confidence: 0.9074827
00:27:34.355 --> 00:27:35.555 well, sometimes this is a
NOTE Confidence: 0.9074827
00:27:35.555 --> 00:27:36.799 Rocky Tanske law. See, this
NOTE Confidence: 0.9074827
00:27:36.799 --> 00:27:37.679 is a picture I took
NOTE Confidence: 0.9074827
00:27:37.679 --> 00:27:39.540 in the eighties in Mexico
NOTE Confidence: 0.9074827
00:27:39.600 --> 00:27:41.040 City where you see the
NOTE Confidence: 0.9074827
00:27:41.040 --> 00:27:42.720 two lungs and then this
NOTE Confidence: 0.9074827
00:27:42.720 --> 00:27:44.480 lobe with a pedicle and
NOTE Confidence: 0.9074827
00:27:44.480 --> 00:27:46.100 vessels. This is a typical
NOTE Confidence: 0.9074827
00:27:46.160 --> 00:27:46.660 sequestration,
NOTE Confidence: 0.9811626
00:27:47.040 --> 00:27:48.240 and we published it with
NOTE Confidence: 0.9811626
00:27:48.240 --> 00:27:49.635 Mark Keller. I don't know
NOTE Confidence: 0.9811626
00:27:49.635 --> 00:27:51.635 if anybody here remembers Mark
NOTE Confidence: 0.9811626

00:27:51.635 --> 00:27:52.915 Keller. He now lives in
NOTE Confidence: 0.9811626

00:27:52.915 --> 00:27:54.515 Miami, close to Miami, and
NOTE Confidence: 0.9811626

00:27:54.515 --> 00:27:55.315 he was the chief of
NOTE Confidence: 0.9811626

00:27:55.315 --> 00:27:56.755 radiology here, and we just
NOTE Confidence: 0.9811626

00:27:56.755 --> 00:27:57.815 had dinner together
NOTE Confidence: 0.95166016

00:27:58.195 --> 00:28:00.055 last week. It's a wonderful
NOTE Confidence: 0.95166016

00:28:00.115 --> 00:28:00.615 colleague.
NOTE Confidence: 0.9763997

00:28:01.075 --> 00:28:02.435 Well, the problem is that
NOTE Confidence: 0.9763997

00:28:02.435 --> 00:28:03.875 many years ago, the group
NOTE Confidence: 0.9763997

00:28:03.875 --> 00:28:04.615 at Harvard,
NOTE Confidence: 0.92820543

00:28:05.290 --> 00:28:06.970 Boston Children's, the h word
NOTE Confidence: 0.92820543

00:28:07.050 --> 00:28:08.330 I'm sorry, John. I remember
NOTE Confidence: 0.92820543

00:28:08.330 --> 00:28:09.630 that the h word is
NOTE Confidence: 0.92820543

00:28:09.850 --> 00:28:11.530 is is problematic here, that
NOTE Confidence: 0.92820543

00:28:11.530 --> 00:28:12.650 evil place of the north
NOTE Confidence: 0.92820543

00:28:12.650 --> 00:28:13.850 as you used to call

NOTE Confidence: 0.92820543

00:28:13.850 --> 00:28:15.050 it, John. But,

NOTE Confidence: 0.8736165

00:28:15.930 --> 00:28:18.190 when the Boston Children's describe

NOTE Confidence: 0.8736165

00:28:18.330 --> 00:28:18.830 this

NOTE Confidence: 0.93322754

00:28:19.425 --> 00:28:20.085 paper, he's

NOTE Confidence: 0.95301104

00:28:20.625 --> 00:28:23.105 they said bronchial atresia is

NOTE Confidence: 0.95301104

00:28:23.105 --> 00:28:24.805 common to extra lower sequestration,

NOTE Confidence: 0.8222692

00:28:25.105 --> 00:28:26.945 intra lower sequestration, and c

NOTE Confidence: 0.8222692

00:28:26.945 --> 00:28:28.645 cam and lower emphysema.

NOTE Confidence: 0.86964923

00:28:29.345 --> 00:28:30.625 So when you obstruct the

NOTE Confidence: 0.86964923

00:28:30.625 --> 00:28:32.405 bronchus in developing

NOTE Confidence: 0.95092773

00:28:32.705 --> 00:28:33.205 kid,

NOTE Confidence: 0.96615833

00:28:33.529 --> 00:28:35.049 that is what happens. Any

NOTE Confidence: 0.96615833

00:28:35.049 --> 00:28:36.090 of those lesions that I

NOTE Confidence: 0.96615833

00:28:36.090 --> 00:28:37.710 mentioned happens. Okay?

NOTE Confidence: 0.99909854

00:28:38.330 --> 00:28:39.450 And there are many papers

NOTE Confidence: 0.99909854

00:28:39.450 --> 00:28:41.450 supporting that. The problem is
NOTE Confidence: 0.99909854

00:28:41.450 --> 00:28:42.350 when you receive
NOTE Confidence: 0.99283856

00:28:42.809 --> 00:28:43.950 something like this.
NOTE Confidence: 0.9370553

00:28:44.570 --> 00:28:45.769 You receive something like this
NOTE Confidence: 0.9370553

00:28:45.769 --> 00:28:46.730 and then you have the
NOTE Confidence: 0.9370553

00:28:46.730 --> 00:28:47.929 cyst here and you say,
NOTE Confidence: 0.9370553

00:28:47.929 --> 00:28:49.425 well, this may be a
NOTE Confidence: 0.9370553

00:28:49.505 --> 00:28:51.505 bronchial atrial deformation sequence or
NOTE Confidence: 0.9370553

00:28:51.505 --> 00:28:52.565 so called CPAM
NOTE Confidence: 0.9013672

00:28:53.025 --> 00:28:54.405 and you see this and
NOTE Confidence: 0.9890408

00:28:55.505 --> 00:28:56.945 you have to consider that
NOTE Confidence: 0.9890408

00:28:56.945 --> 00:28:58.165 this may be a pleural
NOTE Confidence: 0.9890408

00:28:58.225 --> 00:28:59.365 pulmonary blastoma.
NOTE Confidence: 0.9724121

00:29:00.305 --> 00:29:02.085 This is a very aggressive
NOTE Confidence: 0.9724121

00:29:02.145 --> 00:29:03.365 in general tumor
NOTE Confidence: 0.9983724

00:29:03.720 --> 00:29:05.179 that occurs by

NOTE Confidence: 0.9027623
00:29:05.580 --> 00:29:07.620 with mutations of Dicer one.
NOTE Confidence: 0.9027623
00:29:07.640 --> 00:29:08.460 I'm sorry.
NOTE Confidence: 0.9880371
00:29:10.440 --> 00:29:11.419 Dicer one
NOTE Confidence: 0.8284776
00:29:11.720 --> 00:29:13.740 and it occurs in three
NOTE Confidence: 0.8284776
00:29:13.960 --> 00:29:14.460 subtypes,
NOTE Confidence: 0.91590714
00:29:14.840 --> 00:29:16.120 one, two, and three and
NOTE Confidence: 0.91590714
00:29:16.120 --> 00:29:18.040 one is called one, type
NOTE Confidence: 0.91590714
00:29:18.040 --> 00:29:18.860 one regressing.
NOTE Confidence: 0.9087219
00:29:19.534 --> 00:29:20.654 It occurs in the lungs
NOTE Confidence: 0.9087219
00:29:20.654 --> 00:29:21.774 and derives from the long
NOTE Confidence: 0.9087219
00:29:21.774 --> 00:29:23.294 missing kind. But now we
NOTE Confidence: 0.9087219
00:29:23.294 --> 00:29:24.735 are learning that dicer one
NOTE Confidence: 0.9087219
00:29:24.735 --> 00:29:26.355 mutations can produce tumors
NOTE Confidence: 0.9978841
00:29:26.815 --> 00:29:28.595 anywhere and everywhere.
NOTE Confidence: 0.9661187
00:29:28.975 --> 00:29:30.595 The first ones were described
NOTE Confidence: 0.9661187

00:29:30.894 --> 00:29:32.515 in the lung. Okay?
NOTE Confidence: 0.9351671

00:29:33.710 --> 00:29:35.230 They were described by two
NOTE Confidence: 0.9351671

00:29:35.230 --> 00:29:37.230 good friends, Pepper Dehner and
NOTE Confidence: 0.9351671

00:29:37.230 --> 00:29:38.210 Carlos Manibal,
NOTE Confidence: 0.9920044

00:29:39.230 --> 00:29:40.510 who taught me how to
NOTE Confidence: 0.9920044

00:29:40.510 --> 00:29:41.790 do an autopsy for the
NOTE Confidence: 0.9920044

00:29:41.790 --> 00:29:43.950 first time in nineteen eighty
NOTE Confidence: 0.9920044

00:29:43.950 --> 00:29:44.450 six
NOTE Confidence: 0.99239093

00:29:45.165 --> 00:29:46.285 or eighty four. I don't
NOTE Confidence: 0.99239093

00:29:46.285 --> 00:29:46.785 remember.
NOTE Confidence: 0.99072266

00:29:47.565 --> 00:29:48.945 These two guys in Minneapolis,
NOTE Confidence: 0.92450136

00:29:49.565 --> 00:29:50.685 Pepperdiner is one of the
NOTE Confidence: 0.92450136

00:29:50.685 --> 00:29:52.765 most prominent pediatric pathologists in
NOTE Confidence: 0.92450136

00:29:52.765 --> 00:29:53.265 history.
NOTE Confidence: 0.9933456

00:29:53.565 --> 00:29:55.165 And Carlos is a pediatric
NOTE Confidence: 0.9933456

00:29:55.165 --> 00:29:56.605 pathologist at heart, but he's

NOTE Confidence: 0.9933456
00:29:56.605 --> 00:29:57.665 a general pathologist.
NOTE Confidence: 0.9436035
00:29:58.205 --> 00:30:00.220 And they describe this pleural
NOTE Confidence: 0.9436035
00:30:00.220 --> 00:30:01.279 pulmonary blastoma
NOTE Confidence: 0.9995117
00:30:01.820 --> 00:30:02.559 in nineteen
NOTE Confidence: 0.9995117
00:30:02.940 --> 00:30:03.759 eighty eight.
NOTE Confidence: 0.984375
00:30:04.220 --> 00:30:04.720 Well,
NOTE Confidence: 0.9885254
00:30:05.259 --> 00:30:06.159 after that,
NOTE Confidence: 0.7659058
00:30:06.860 --> 00:30:09.360 Ashley Hill, Jason Hor, Jason
NOTE Confidence: 0.7659058
00:30:09.500 --> 00:30:10.000 Jarsemboski
NOTE Confidence: 0.8149414
00:30:10.460 --> 00:30:11.440 and Pepper Dehner
NOTE Confidence: 0.9041341
00:30:11.820 --> 00:30:13.980 describe the mutations on these
NOTE Confidence: 0.9041341
00:30:13.980 --> 00:30:14.475 lesions
NOTE Confidence: 0.9439697
00:30:14.955 --> 00:30:15.595 and then,
NOTE Confidence: 0.9366319
00:30:16.315 --> 00:30:18.395 identify the major genetic factor
NOTE Confidence: 0.9366319
00:30:18.395 --> 00:30:19.855 which is Dicer one
NOTE Confidence: 0.9671631

00:30:20.315 --> 00:30:21.914 mutations that lead to the
NOTE Confidence: 0.9671631

00:30:21.914 --> 00:30:23.215 Dicer one syndrome.
NOTE Confidence: 0.82714844

00:30:23.835 --> 00:30:24.335 These,
NOTE Confidence: 0.8327637

00:30:24.955 --> 00:30:25.455 are,
NOTE Confidence: 0.9601863

00:30:26.075 --> 00:30:27.929 mutations that affect a key
NOTE Confidence: 0.9601863

00:30:27.929 --> 00:30:29.950 enzyme required to keep precursors
NOTE Confidence: 0.9601863

00:30:30.010 --> 00:30:31.470 of micro RNAs
NOTE Confidence: 0.9451904

00:30:32.090 --> 00:30:34.429 into their mature active forms.
NOTE Confidence: 0.9451904

00:30:34.649 --> 00:30:35.950 And when they are deleted,
NOTE Confidence: 0.92301434

00:30:36.730 --> 00:30:38.269 they are, very,
NOTE Confidence: 0.9864909

00:30:39.289 --> 00:30:40.429 severe in effects.
NOTE Confidence: 0.6069336

00:30:41.065 --> 00:30:41.565 The,
NOTE Confidence: 0.82698566

00:30:41.945 --> 00:30:42.445 germline,
NOTE Confidence: 0.99902344

00:30:42.985 --> 00:30:43.485 pathogenic
NOTE Confidence: 0.84231424

00:30:43.865 --> 00:30:45.545 Dicer one variants, they find
NOTE Confidence: 0.84231424

00:30:45.545 --> 00:30:46.285 these syndromes

NOTE Confidence: 0.94873047

00:30:46.745 --> 00:30:48.745 are always loss of function

NOTE Confidence: 0.94873047

00:30:48.745 --> 00:30:49.245 type

NOTE Confidence: 0.9336495

00:30:49.625 --> 00:30:51.225 and, like many other tumor

NOTE Confidence: 0.9336495

00:30:51.225 --> 00:30:53.945 suppressor genes, germline, allelic loss

NOTE Confidence: 0.9336495

00:30:53.945 --> 00:30:55.465 of function mutations in Dicer

NOTE Confidence: 0.9336495

00:30:55.465 --> 00:30:57.399 one create tumor susceptibility,

NOTE Confidence: 0.961497

00:30:57.940 --> 00:30:59.700 but appear to be insufficient

NOTE Confidence: 0.961497

00:30:59.700 --> 00:31:01.080 to initiate tumorigenesis.

NOTE Confidence: 0.9342041

00:31:01.700 --> 00:31:03.620 There are other changes required

NOTE Confidence: 0.9342041

00:31:03.620 --> 00:31:04.679 to develop into,

NOTE Confidence: 0.9637451

00:31:05.299 --> 00:31:05.799 tumorigenesis.

NOTE Confidence: 0.9977539

00:31:06.659 --> 00:31:08.440 Many different organs are affected.

NOTE Confidence: 0.9134776

00:31:08.875 --> 00:31:10.075 The first one described was

NOTE Confidence: 0.9134776

00:31:10.075 --> 00:31:10.575 pulopulmonary

NOTE Confidence: 0.8468933

00:31:10.955 --> 00:31:12.395 blastoma, but we now know

NOTE Confidence: 0.8468933

00:31:12.395 --> 00:31:14.235 that cystic nephroma and a
NOTE Confidence: 0.8468933

00:31:14.235 --> 00:31:15.934 plastic sarcoma of the kidney,
NOTE Confidence: 0.8468933

00:31:16.154 --> 00:31:17.375 and brianal rhabdomyosarcomas
NOTE Confidence: 0.7672526

00:31:18.154 --> 00:31:19.775 and cervical embryonal
NOTE Confidence: 0.92285156

00:31:20.315 --> 00:31:20.815 rhabdomyosarcomas
NOTE Confidence: 0.9302146

00:31:21.755 --> 00:31:23.355 are also associated with Dicer
NOTE Confidence: 0.9302146

00:31:23.355 --> 00:31:25.100 one and many other types
NOTE Confidence: 0.9302146

00:31:25.100 --> 00:31:27.500 of tumors including endocrine, CNS
NOTE Confidence: 0.9302146

00:31:27.500 --> 00:31:30.059 tumors, and ovarian tumors, gyn,
NOTE Confidence: 0.9302146

00:31:30.059 --> 00:31:30.559 gynandroblastoma.
NOTE Confidence: 0.9841919

00:31:31.899 --> 00:31:33.100 This is an example of
NOTE Confidence: 0.9841919

00:31:33.100 --> 00:31:34.240 a Dicer one,
NOTE Confidence: 0.9941406

00:31:34.779 --> 00:31:35.279 tumor.
NOTE Confidence: 0.9349365

00:31:36.139 --> 00:31:37.019 You can see is,
NOTE Confidence: 0.9716797

00:31:37.755 --> 00:31:39.435 one of the cystic types
NOTE Confidence: 0.9716797

00:31:39.435 --> 00:31:40.955 not as aggressive but they

NOTE Confidence: 0.9716797

00:31:40.955 --> 00:31:42.655 can be like this

NOTE Confidence: 0.94162595

00:31:43.595 --> 00:31:45.675 or like this or like

NOTE Confidence: 0.94162595

00:31:45.675 --> 00:31:47.995 this, pleural pulmonary blastoma cystic

NOTE Confidence: 0.94162595

00:31:47.995 --> 00:31:49.695 that can be very cystic.

NOTE Confidence: 0.9585067

00:31:50.560 --> 00:31:51.760 This is very difficult to

NOTE Confidence: 0.9585067

00:31:51.760 --> 00:31:53.360 diagnose as a pleural pulmonary

NOTE Confidence: 0.9585067

00:31:53.360 --> 00:31:54.960 blastoma because it looks completely

NOTE Confidence: 0.9585067

00:31:54.960 --> 00:31:56.960 benign and it behaves in

NOTE Confidence: 0.9585067

00:31:56.960 --> 00:31:58.020 a benign fashion.

NOTE Confidence: 0.99465764

00:31:58.480 --> 00:31:59.940 But you have to identify

NOTE Confidence: 0.99465764

00:32:00.000 --> 00:32:01.760 because the patient and the

NOTE Confidence: 0.99465764

00:32:01.760 --> 00:32:03.760 patient's family is at risk

NOTE Confidence: 0.99465764

00:32:03.760 --> 00:32:04.500 of developing

NOTE Confidence: 0.9970703

00:32:04.800 --> 00:32:05.700 other tumors.

NOTE Confidence: 0.956208

00:32:08.065 --> 00:32:09.365 Here you have a repetition

NOTE Confidence: 0.956208

00:32:09.424 --> 00:32:10.304 of what I just said.
NOTE Confidence: 0.956208

00:32:10.304 --> 00:32:11.505 I'm not gonna spend more
NOTE Confidence: 0.956208

00:32:11.505 --> 00:32:12.785 time here. This is how
NOTE Confidence: 0.956208

00:32:12.785 --> 00:32:13.985 you pay attention to the
NOTE Confidence: 0.956208

00:32:13.985 --> 00:32:16.225 stroma and diagnose it as
NOTE Confidence: 0.956208

00:32:16.225 --> 00:32:18.245 a plural pleural pulmonary blastoma
NOTE Confidence: 0.956208

00:32:18.304 --> 00:32:20.164 solid with rhabdomyoblastoma
NOTE Confidence: 0.98706055

00:32:20.945 --> 00:32:22.160 like elements
NOTE Confidence: 0.80050784

00:32:22.460 --> 00:32:24.480 with an aplasia, abundant cytoplasm.
NOTE Confidence: 0.95806885

00:32:25.020 --> 00:32:26.060 And then when you proceed
NOTE Confidence: 0.95806885

00:32:26.060 --> 00:32:27.580 with the genetic analysis, you
NOTE Confidence: 0.95806885

00:32:27.580 --> 00:32:29.580 identify the germline Dicer one
NOTE Confidence: 0.95806885

00:32:29.580 --> 00:32:30.080 mutations
NOTE Confidence: 0.9851074

00:32:30.620 --> 00:32:31.100 in,
NOTE Confidence: 0.7902832

00:32:31.500 --> 00:32:32.000 these,
NOTE Confidence: 0.82299805

00:32:33.180 --> 00:32:33.680 tissues.

NOTE Confidence: 0.9156494
00:32:34.285 --> 00:32:35.645 You can also use,
NOTE Confidence: 0.9433594
00:32:36.125 --> 00:32:36.625 immunohistochemistry,
NOTE Confidence: 1
00:32:37.965 --> 00:32:38.465 for
NOTE Confidence: 0.8453776
00:32:39.565 --> 00:32:40.945 labeling these changes.
NOTE Confidence: 0.9416822
00:32:41.325 --> 00:32:42.125 Let me show you a
NOTE Confidence: 0.9416822
00:32:42.125 --> 00:32:43.405 council that I received a
NOTE Confidence: 0.9416822
00:32:43.405 --> 00:32:45.005 few years ago. Came from
NOTE Confidence: 0.9416822
00:32:45.005 --> 00:32:46.605 Italy in a patient of
NOTE Confidence: 0.9416822
00:32:46.605 --> 00:32:47.745 one year old
NOTE Confidence: 0.9562988
00:32:48.560 --> 00:32:49.060 with,
NOTE Confidence: 0.97229004
00:32:49.640 --> 00:32:50.480 I'm sure all of you
NOTE Confidence: 0.97229004
00:32:50.480 --> 00:32:51.460 can read Italian.
NOTE Confidence: 0.9671097
00:32:52.160 --> 00:32:54.100 The patient had medulloepithelioma
NOTE Confidence: 0.98917645
00:32:55.040 --> 00:32:55.940 in the eye,
NOTE Confidence: 0.9770508
00:32:56.400 --> 00:32:56.900 retinoblastoma
NOTE Confidence: 0.7026367

00:32:58.080 --> 00:32:58.480 and,
NOTE Confidence: 0.7911682
00:32:59.440 --> 00:33:00.580 on the one enucleation.
NOTE Confidence: 0.9785889
00:33:00.960 --> 00:33:02.735 So the patient already had
NOTE Confidence: 0.9785889
00:33:02.735 --> 00:33:04.115 a tumor in the eye,
NOTE Confidence: 0.8414714
00:33:04.575 --> 00:33:05.075 retinoblastoma.
NOTE Confidence: 0.92380595
00:33:05.534 --> 00:33:07.154 It's a relatively common pediatric
NOTE Confidence: 0.92380595
00:33:07.215 --> 00:33:09.375 tumor and then the patient
NOTE Confidence: 0.92380595
00:33:09.375 --> 00:33:09.875 has
NOTE Confidence: 0.9943034
00:33:10.654 --> 00:33:11.794 a long lesion
NOTE Confidence: 0.94383407
00:33:12.174 --> 00:33:13.475 and this is the appearance
NOTE Confidence: 0.94383407
00:33:13.534 --> 00:33:14.975 of the long lesion that
NOTE Confidence: 0.94383407
00:33:14.975 --> 00:33:16.414 I received. These are images
NOTE Confidence: 0.94383407
00:33:16.414 --> 00:33:18.009 taken from that,
NOTE Confidence: 0.91813153
00:33:18.549 --> 00:33:19.029 that's,
NOTE Confidence: 0.9970703
00:33:19.350 --> 00:33:19.850 case
NOTE Confidence: 0.89767635
00:33:20.230 --> 00:33:21.590 and as you appreciate this

NOTE Confidence: 0.89767635
00:33:21.590 --> 00:33:22.950 cyst and you pay attention
NOTE Confidence: 0.89767635
00:33:22.950 --> 00:33:24.549 to the stroma component of
NOTE Confidence: 0.89767635
00:33:24.549 --> 00:33:25.289 this cyst,
NOTE Confidence: 0.99560547
00:33:26.070 --> 00:33:27.690 something is not right
NOTE Confidence: 0.9511719
00:33:27.990 --> 00:33:28.950 and then you go in
NOTE Confidence: 0.9511719
00:33:28.950 --> 00:33:29.690 other areas
NOTE Confidence: 0.8897705
00:33:30.470 --> 00:33:32.250 more cellular and then
NOTE Confidence: 0.9061937
00:33:32.825 --> 00:33:34.525 the diagnosis that we issued
NOTE Confidence: 0.9061937
00:33:34.585 --> 00:33:36.285 was this type one cystic
NOTE Confidence: 0.9061937
00:33:36.425 --> 00:33:37.725 pure pulmonary blastoma.
NOTE Confidence: 0.9533343
00:33:38.025 --> 00:33:39.065 We didn't know about the
NOTE Confidence: 0.9533343
00:33:39.065 --> 00:33:40.905 resection margins. We said that
NOTE Confidence: 0.9533343
00:33:40.905 --> 00:33:42.025 there was no involvement of
NOTE Confidence: 0.9533343
00:33:42.025 --> 00:33:43.225 the plural in the material
NOTE Confidence: 0.9533343
00:33:43.225 --> 00:33:43.725 reviewed
NOTE Confidence: 0.886792

00:33:44.105 --> 00:33:46.205 and the background long parenchyma
NOTE Confidence: 0.9259033

00:33:46.585 --> 00:33:47.645 looked normal.
NOTE Confidence: 0.92210627

00:33:48.480 --> 00:33:49.840 And we made a comment
NOTE Confidence: 0.92210627

00:33:49.840 --> 00:33:51.360 here and referred to the
NOTE Confidence: 0.92210627

00:33:51.360 --> 00:33:53.360 pure, to the beats five
NOTE Confidence: 0.92210627

00:33:53.360 --> 00:33:54.820 edition of the pediatric
NOTE Confidence: 0.99869794

00:33:55.600 --> 00:33:56.580 tumors in the
NOTE Confidence: 0.9899514

00:33:58.240 --> 00:33:59.360 WHO. Let me show you
NOTE Confidence: 0.9899514

00:33:59.360 --> 00:34:00.720 another example. This is a
NOTE Confidence: 0.9899514

00:34:00.720 --> 00:34:02.075 two year old with a
NOTE Confidence: 0.9899514

00:34:02.075 --> 00:34:03.755 left pneumothorax that you can
NOTE Confidence: 0.9899514

00:34:03.755 --> 00:34:04.735 appreciate here
NOTE Confidence: 0.7067871

00:34:05.115 --> 00:34:05.775 and then
NOTE Confidence: 0.9567871

00:34:06.555 --> 00:34:07.915 similar lesions to the one
NOTE Confidence: 0.9567871

00:34:07.915 --> 00:34:09.135 that I just discussed.
NOTE Confidence: 0.98916477

00:34:09.995 --> 00:34:12.975 They resected these lesions. Unfortunately,

NOTE Confidence: 0.98916477
00:34:13.195 --> 00:34:13.935 the specimen
NOTE Confidence: 0.9569092
00:34:14.235 --> 00:34:16.495 perforated and it deflated.
NOTE Confidence: 0.96867317
00:34:16.800 --> 00:34:18.340 It was a cystic lesion
NOTE Confidence: 0.96867317
00:34:18.560 --> 00:34:20.880 that appears collapsed here. This
NOTE Confidence: 0.96867317
00:34:20.880 --> 00:34:22.480 is the ink that, our,
NOTE Confidence: 0.96867317
00:34:22.880 --> 00:34:24.260 residents or PAs,
NOTE Confidence: 0.94295245
00:34:25.040 --> 00:34:26.580 dissecting this, example
NOTE Confidence: 0.950486
00:34:26.960 --> 00:34:28.480 put on to, evaluate the
NOTE Confidence: 0.950486
00:34:28.480 --> 00:34:30.000 margins. And then when we
NOTE Confidence: 0.950486
00:34:30.000 --> 00:34:30.980 look at the histology,
NOTE Confidence: 0.9700185
00:34:31.594 --> 00:34:33.114 there are areas like normal
NOTE Confidence: 0.9700185
00:34:33.114 --> 00:34:35.454 lung, areas with little cysts,
NOTE Confidence: 0.9700185
00:34:35.594 --> 00:34:36.714 but then there are areas
NOTE Confidence: 0.9700185
00:34:36.714 --> 00:34:37.535 like this
NOTE Confidence: 0.8465576
00:34:38.954 --> 00:34:39.775 and this.
NOTE Confidence: 0.9515991

00:34:40.075 --> 00:34:41.614 So this is not,
NOTE Confidence: 0.92929417
00:34:43.640 --> 00:34:45.239 as formerly referred to a
NOTE Confidence: 0.92929417
00:34:45.239 --> 00:34:46.760 CPAM. This is a pleural
NOTE Confidence: 0.92929417
00:34:46.760 --> 00:34:47.739 pulmonary blastoma,
NOTE Confidence: 0.9780816
00:34:48.040 --> 00:34:49.080 and you have to pay
NOTE Confidence: 0.9780816
00:34:49.080 --> 00:34:50.380 attention to this because
NOTE Confidence: 0.9956752
00:34:50.680 --> 00:34:51.960 that is what is going
NOTE Confidence: 0.9956752
00:34:51.960 --> 00:34:52.620 to determine
NOTE Confidence: 0.9823843
00:34:52.920 --> 00:34:54.360 the progression of the disease
NOTE Confidence: 0.9823843
00:34:54.360 --> 00:34:56.200 and the treatment necessary to
NOTE Confidence: 0.9823843
00:34:56.200 --> 00:34:57.255 control this entity.
NOTE Confidence: 0.9184672
00:34:57.654 --> 00:34:59.094 There is an aplasia in
NOTE Confidence: 0.9184672
00:34:59.094 --> 00:35:00.394 this stromal cells,
NOTE Confidence: 0.9275553
00:35:00.694 --> 00:35:02.454 very significant an aplasia in
NOTE Confidence: 0.9275553
00:35:02.454 --> 00:35:03.974 some areas, and we issued
NOTE Confidence: 0.9275553
00:35:03.974 --> 00:35:05.734 a diagnosis. Actually, this was

NOTE Confidence: 0.9275553

00:35:05.734 --> 00:35:06.694 the case that there was

NOTE Confidence: 0.9275553

00:35:06.694 --> 00:35:07.894 not mine, but it was

NOTE Confidence: 0.9275553

00:35:07.894 --> 00:35:09.015 in my department. And the,

NOTE Confidence: 0.9436384

00:35:09.969 --> 00:35:11.489 the all these cases were,

NOTE Confidence: 0.9436384

00:35:11.730 --> 00:35:12.930 shared with us and they

NOTE Confidence: 0.9436384

00:35:12.930 --> 00:35:14.070 they used to incorporate

NOTE Confidence: 0.8795573

00:35:14.450 --> 00:35:16.150 our opinions on the pathology

NOTE Confidence: 0.8795573

00:35:16.290 --> 00:35:17.890 signing this case, call it

NOTE Confidence: 0.8795573

00:35:17.890 --> 00:35:19.969 pluripronary blastoma type two because

NOTE Confidence: 0.8795573

00:35:19.969 --> 00:35:21.910 it's not the very cystic

NOTE Confidence: 0.8795573

00:35:22.205 --> 00:35:23.485 but not the very solid.

NOTE Confidence: 0.8795573

00:35:23.485 --> 00:35:24.305 It's mixed

NOTE Confidence: 0.7504883

00:35:24.685 --> 00:35:25.265 and then

NOTE Confidence: 0.6185826

00:35:25.645 --> 00:35:27.725 multiple multifocal an aplasia, a

NOTE Confidence: 0.6185826

00:35:27.725 --> 00:35:28.625 random myometres,

NOTE Confidence: 0.91186523

00:35:29.245 --> 00:35:31.425 differentiation with multifocal an aplasia,
NOTE Confidence: 0.91186523

00:35:31.645 --> 00:35:33.405 etcetera, etcetera, and they comment
NOTE Confidence: 0.91186523

00:35:33.405 --> 00:35:33.905 here.
NOTE Confidence: 0.9917806

00:35:38.150 --> 00:35:39.050 How about this?
NOTE Confidence: 0.9925537

00:35:39.750 --> 00:35:40.550 We are not in the
NOTE Confidence: 0.9925537

00:35:40.550 --> 00:35:41.050 lung.
NOTE Confidence: 0.9775065

00:35:41.750 --> 00:35:43.450 This is a cystic lesion.
NOTE Confidence: 0.93811035

00:35:44.550 --> 00:35:46.170 You recognize this. Right?
NOTE Confidence: 0.88757324

00:35:47.110 --> 00:35:47.930 The testis
NOTE Confidence: 0.863739

00:35:48.470 --> 00:35:49.530 is in the paratesticular
NOTE Confidence: 0.99902344

00:35:50.550 --> 00:35:51.050 area.
NOTE Confidence: 1

00:35:51.835 --> 00:35:52.495 I put
NOTE Confidence: 0.95377606

00:35:53.195 --> 00:35:54.795 my cell phone under the
NOTE Confidence: 0.95377606

00:35:54.795 --> 00:35:56.575 glass taking the picture
NOTE Confidence: 0.9729614

00:35:56.875 --> 00:35:58.735 to see the transillumination
NOTE Confidence: 0.99869794

00:35:59.515 --> 00:36:00.415 of the cyst.

NOTE Confidence: 0.99560547
00:36:00.795 --> 00:36:01.295 Okay?
NOTE Confidence: 0.9803738
00:36:01.835 --> 00:36:03.435 And, yeah, residents laugh at
NOTE Confidence: 0.9803738
00:36:03.435 --> 00:36:04.315 me when I do these
NOTE Confidence: 0.9803738
00:36:04.315 --> 00:36:06.150 kinds of things because they
NOTE Confidence: 0.9803738
00:36:06.150 --> 00:36:07.190 say, why did you do
NOTE Confidence: 0.9803738
00:36:07.190 --> 00:36:08.390 that? Well, I want to
NOTE Confidence: 0.9803738
00:36:08.390 --> 00:36:09.829 see the trans illumination. Right?
NOTE Confidence: 0.9803738
00:36:09.829 --> 00:36:10.710 And and you just have
NOTE Confidence: 0.9803738
00:36:10.710 --> 00:36:11.210 to
NOTE Confidence: 0.9973958
00:36:11.910 --> 00:36:13.050 follow your imagination.
NOTE Confidence: 0.9944661
00:36:13.829 --> 00:36:15.670 And, when when I did
NOTE Confidence: 0.9944661
00:36:15.670 --> 00:36:16.170 this,
NOTE Confidence: 0.9248589
00:36:16.950 --> 00:36:18.905 the fellow that was rotating
NOTE Confidence: 0.9248589
00:36:18.965 --> 00:36:20.325 and doing his fellowship with
NOTE Confidence: 0.9248589
00:36:20.325 --> 00:36:21.844 us in Pittsburgh, doctor Louis
NOTE Confidence: 0.9248589

00:36:21.844 --> 00:36:23.305 Samson, a fantastic,
NOTE Confidence: 0.8919678

00:36:23.685 --> 00:36:25.844 fellow pediatric pathology fellow from
NOTE Confidence: 0.8919678

00:36:25.844 --> 00:36:26.344 Canada,
NOTE Confidence: 0.90999347

00:36:26.805 --> 00:36:27.685 he put this,
NOTE Confidence: 0.9728782

00:36:29.285 --> 00:36:30.565 pictures in the tumor board,
NOTE Confidence: 0.9728782

00:36:30.565 --> 00:36:31.685 and this was the final
NOTE Confidence: 0.9728782

00:36:31.685 --> 00:36:32.185 diagnosis.
NOTE Confidence: 0.9264729

00:36:32.520 --> 00:36:35.480 Cystically dilated epididymal ducts consisting
NOTE Confidence: 0.9264729

00:36:35.480 --> 00:36:37.160 with obstructive changes. Yes. Yes.
NOTE Confidence: 0.9264729

00:36:37.160 --> 00:36:38.680 Yes. And then low grade
NOTE Confidence: 0.9264729

00:36:38.680 --> 00:36:40.219 myxoid spin cell neoplasm
NOTE Confidence: 0.9836426

00:36:40.680 --> 00:36:41.580 with a comment.
NOTE Confidence: 0.9647217

00:36:41.880 --> 00:36:43.160 And we learned that in
NOTE Confidence: 0.9647217

00:36:43.160 --> 00:36:45.020 the clinical history, there was
NOTE Confidence: 0.8843587

00:36:45.320 --> 00:36:46.920 a history of Dicer one
NOTE Confidence: 0.8843587

00:36:46.920 --> 00:36:47.420 syndrome.

NOTE Confidence: 0.98413086
00:36:48.015 --> 00:36:49.954 So this lesion fits perfectly
NOTE Confidence: 0.98413086
00:36:50.015 --> 00:36:50.515 well.
NOTE Confidence: 0.99964195
00:36:51.135 --> 00:36:52.255 The fact that we saw
NOTE Confidence: 0.99964195
00:36:52.255 --> 00:36:53.694 it after the patient has
NOTE Confidence: 0.99964195
00:36:53.694 --> 00:36:55.395 been diagnosed with Dicer one
NOTE Confidence: 0.9669667
00:36:55.775 --> 00:36:57.214 is good, but it doesn't
NOTE Confidence: 0.9669667
00:36:57.214 --> 00:36:59.055 matter. It fits everything. And
NOTE Confidence: 0.9669667
00:36:59.055 --> 00:37:00.414 if you see this before
NOTE Confidence: 0.9669667
00:37:00.414 --> 00:37:02.255 knowing the clinical history, you
NOTE Confidence: 0.9669667
00:37:02.255 --> 00:37:03.555 are after it.
NOTE Confidence: 0.73876953
00:37:05.080 --> 00:37:06.219 Recently, group
NOTE Confidence: 0.9765625
00:37:06.520 --> 00:37:07.020 in,
NOTE Confidence: 0.9750163
00:37:07.800 --> 00:37:08.920 that evil place of the
NOTE Confidence: 0.9750163
00:37:08.920 --> 00:37:09.420 north,
NOTE Confidence: 0.8887695
00:37:09.880 --> 00:37:11.580 published this series of cases
NOTE Confidence: 0.8104553

00:37:11.960 --> 00:37:13.820 with Dicer one related
NOTE Confidence: 0.8913574

00:37:14.200 --> 00:37:15.660 WIMS like uterine
NOTE Confidence: 0.7573242

00:37:16.040 --> 00:37:16.540 tumor,
NOTE Confidence: 0.9843913

00:37:17.385 --> 00:37:19.305 with, several cases reported. And
NOTE Confidence: 0.9843913

00:37:19.305 --> 00:37:21.325 the the series are growing
NOTE Confidence: 0.9843913

00:37:21.465 --> 00:37:23.325 in adults and in pediatric
NOTE Confidence: 0.9181641

00:37:23.705 --> 00:37:25.885 examples of Dicer one mutations.
NOTE Confidence: 0.99869794

00:37:28.839 --> 00:37:30.700 Let me switch to another
NOTE Confidence: 0.99869794

00:37:30.839 --> 00:37:31.339 interesting
NOTE Confidence: 1

00:37:31.719 --> 00:37:32.219 finding.
NOTE Confidence: 0.9139752

00:37:32.680 --> 00:37:33.640 This is a five year
NOTE Confidence: 0.9139752

00:37:33.640 --> 00:37:35.319 old girl with a palpable
NOTE Confidence: 0.9139752

00:37:35.319 --> 00:37:36.599 or propopubic mass. This is
NOTE Confidence: 0.9139752

00:37:36.599 --> 00:37:37.579 a Yale case
NOTE Confidence: 0.9676888

00:37:38.520 --> 00:37:40.200 that, came to us with
NOTE Confidence: 0.9676888

00:37:40.200 --> 00:37:42.565 two independent masses. And, when

NOTE Confidence: 0.9676888

00:37:42.565 --> 00:37:43.525 I was looking at this

NOTE Confidence: 0.9676888

00:37:43.525 --> 00:37:45.065 case, I said two masses.

NOTE Confidence: 0.9676888

00:37:45.205 --> 00:37:46.825 And I remember doctor McNamara,

NOTE Confidence: 0.97867835

00:37:47.285 --> 00:37:48.105 Joe McNamara,

NOTE Confidence: 0.97712404

00:37:48.405 --> 00:37:49.864 one of the pediatric oncologist,

NOTE Confidence: 0.92137194

00:37:50.725 --> 00:37:52.645 told me, yeah, our surgeons

NOTE Confidence: 0.92137194

00:37:52.645 --> 00:37:53.765 think that this is a

NOTE Confidence: 0.92137194

00:37:53.765 --> 00:37:55.445 Williams tumor that dropped down

NOTE Confidence: 0.92137194

00:37:55.445 --> 00:37:57.150 a metastasis into the bladder.

NOTE Confidence: 0.92137194

00:37:57.150 --> 00:37:58.609 I had never seen that.

NOTE Confidence: 0.92137194

00:37:58.670 --> 00:37:59.789 And I have not seen

NOTE Confidence: 0.92137194

00:37:59.789 --> 00:38:01.250 that still now.

NOTE Confidence: 0.9298619

00:38:02.109 --> 00:38:03.230 So five year old with

NOTE Confidence: 0.9298619

00:38:03.230 --> 00:38:03.890 two tumors,

NOTE Confidence: 0.9886882

00:38:04.190 --> 00:38:05.569 there was, however,

NOTE Confidence: 0.9970703

00:38:06.670 --> 00:38:07.170 developmental
NOTE Confidence: 0.9779297
00:38:07.469 --> 00:38:09.089 delay in the clinical history.
NOTE Confidence: 0.9975586
00:38:09.630 --> 00:38:11.010 And because of that
NOTE Confidence: 0.9645996
00:38:11.575 --> 00:38:12.395 and facial
NOTE Confidence: 0.8255615
00:38:13.175 --> 00:38:13.675 dysmorphism,
NOTE Confidence: 0.982137
00:38:14.614 --> 00:38:16.315 a karyotype was obtained
NOTE Confidence: 0.9679314
00:38:16.695 --> 00:38:18.135 at that time, and they
NOTE Confidence: 0.9679314
00:38:18.135 --> 00:38:20.055 found an interstitial deletion of
NOTE Confidence: 0.9679314
00:38:20.055 --> 00:38:21.815 nine q twenty two q
NOTE Confidence: 0.9679314
00:38:21.815 --> 00:38:22.635 thirty two.
NOTE Confidence: 0.9965069
00:38:23.750 --> 00:38:24.790 And that part of the
NOTE Confidence: 0.9965069
00:38:24.790 --> 00:38:26.650 clinical history made me think
NOTE Confidence: 0.9965069
00:38:26.710 --> 00:38:27.770 in something else.
NOTE Confidence: 0.9693554
00:38:29.510 --> 00:38:30.869 In addition, there was mild
NOTE Confidence: 0.9693554
00:38:30.869 --> 00:38:32.089 hypertrophy, hemihypertrophy,
NOTE Confidence: 0.99853516
00:38:32.630 --> 00:38:33.609 prominent forehead,

NOTE Confidence: 0.9532572
00:38:34.150 --> 00:38:36.309 hyper telierism, epicanthal folds, etcetera,
NOTE Confidence: 0.9532572
00:38:36.309 --> 00:38:37.750 etcetera, etcetera, all that you
NOTE Confidence: 0.9532572
00:38:37.750 --> 00:38:39.414 read there. These are the
NOTE Confidence: 0.9532572
00:38:39.414 --> 00:38:41.275 images of the kidney tumor
NOTE Confidence: 0.96777344
00:38:41.974 --> 00:38:42.474 with
NOTE Confidence: 0.8063965
00:38:43.094 --> 00:38:43.835 a epithelial
NOTE Confidence: 0.67858887
00:38:44.295 --> 00:38:44.795 slash,
NOTE Confidence: 0.7288411
00:38:46.135 --> 00:38:46.635 blastema,
NOTE Confidence: 0.90865386
00:38:47.494 --> 00:38:49.335 components in the Wilms tumor
NOTE Confidence: 0.90865386
00:38:49.335 --> 00:38:50.535 that looks a little weird,
NOTE Confidence: 0.90865386
00:38:50.535 --> 00:38:51.594 but it is
NOTE Confidence: 0.8572591
00:38:51.895 --> 00:38:53.035 a Wilms tumor.
NOTE Confidence: 0.99273
00:38:53.940 --> 00:38:55.300 And these are the images
NOTE Confidence: 0.99273
00:38:55.300 --> 00:38:56.520 of the bladder too.
NOTE Confidence: 0.99902344
00:38:57.140 --> 00:38:58.520 If you see them separately,
NOTE Confidence: 0.9201416

00:38:58.900 --> 00:39:00.520 you will diagnose Wim's tumor
NOTE Confidence: 0.9437968

00:39:00.900 --> 00:39:02.660 next, and you will diagnose
NOTE Confidence: 0.9437968

00:39:02.660 --> 00:39:03.160 rhabdomyosarcoma
NOTE Confidence: 0.9711914

00:39:04.260 --> 00:39:04.760 next.
NOTE Confidence: 0.9171491

00:39:05.380 --> 00:39:07.380 Here, we have stroma with
NOTE Confidence: 0.9171491

00:39:07.380 --> 00:39:08.119 an aplasia,
NOTE Confidence: 0.7483724

00:39:08.934 --> 00:39:09.434 desmin,
NOTE Confidence: 0.8434143

00:39:10.535 --> 00:39:12.555 myogenic expression, classic rhabdomyosarcoma.
NOTE Confidence: 0.9477539

00:39:13.654 --> 00:39:14.694 But the patient has two
NOTE Confidence: 0.9477539

00:39:14.694 --> 00:39:15.895 tumors and is five years
NOTE Confidence: 0.9477539

00:39:15.895 --> 00:39:16.474 of age.
NOTE Confidence: 0.9913505

00:39:17.015 --> 00:39:18.055 If you see that in
NOTE Confidence: 0.9913505

00:39:18.055 --> 00:39:18.714 a sixty
NOTE Confidence: 0.99161786

00:39:19.255 --> 00:39:21.255 something years old patient, two
NOTE Confidence: 0.99161786

00:39:21.255 --> 00:39:21.755 tumors
NOTE Confidence: 0.95198566

00:39:22.214 --> 00:39:23.515 is very common

NOTE Confidence: 0.90515137
00:39:23.870 --> 00:39:24.770 in a kid?
NOTE Confidence: 0.99560547
00:39:25.150 --> 00:39:25.650 No.
NOTE Confidence: 0.9570887
00:39:27.469 --> 00:39:29.150 So when I re review
NOTE Confidence: 0.9570887
00:39:29.150 --> 00:39:30.750 the clinical history and I
NOTE Confidence: 0.9570887
00:39:30.750 --> 00:39:32.590 saw the interstitial deletion in
NOTE Confidence: 0.9570887
00:39:32.590 --> 00:39:33.650 chromosome nine,
NOTE Confidence: 0.9995117
00:39:34.030 --> 00:39:35.710 I went to my late
NOTE Confidence: 0.9995117
00:39:35.710 --> 00:39:36.210 wife's
NOTE Confidence: 0.99785155
00:39:36.635 --> 00:39:38.575 lab in the genetics department
NOTE Confidence: 0.88427734
00:39:38.875 --> 00:39:40.734 working with doctor Alan Bale.
NOTE Confidence: 0.96835935
00:39:41.594 --> 00:39:43.295 Myra was my late wife,
NOTE Confidence: 0.9764333
00:39:43.835 --> 00:39:45.194 and she was working in
NOTE Confidence: 0.9764333
00:39:45.194 --> 00:39:47.194 that laboratory. And Alan Bale
NOTE Confidence: 0.9764333
00:39:47.194 --> 00:39:48.315 happens to be the person
NOTE Confidence: 0.9764333
00:39:48.315 --> 00:39:49.055 that identify
NOTE Confidence: 0.9980469

00:39:49.675 --> 00:39:50.415 the patched
NOTE Confidence: 0.91430664

00:39:50.875 --> 00:39:51.375 gene.
NOTE Confidence: 0.9977214

00:39:52.390 --> 00:39:53.369 And the patched
NOTE Confidence: 0.9864909

00:39:53.670 --> 00:39:54.810 gene is responsible
NOTE Confidence: 0.6786194

00:39:55.350 --> 00:39:56.650 for Goling
NOTE Confidence: 0.9069824

00:39:57.030 --> 00:39:57.530 syndrome,
NOTE Confidence: 0.99853516

00:39:58.070 --> 00:39:59.690 which is a cancer predisposition
NOTE Confidence: 0.84521484

00:40:00.070 --> 00:40:00.570 syndrome,
NOTE Confidence: 0.996521

00:40:01.030 --> 00:40:02.890 a very important cancer predisposition
NOTE Confidence: 0.996521

00:40:03.110 --> 00:40:04.650 syndrome. You can
NOTE Confidence: 0.91878796

00:40:05.625 --> 00:40:07.225 advise the clinicians to pursue
NOTE Confidence: 0.91878796

00:40:07.225 --> 00:40:08.905 the diagnosis of Golgi syndrome
NOTE Confidence: 0.91878796

00:40:08.905 --> 00:40:10.205 when you receive an odontogenic
NOTE Confidence: 0.9325684

00:40:10.585 --> 00:40:12.925 keratocyst because they are commonly
NOTE Confidence: 0.9983724

00:40:13.385 --> 00:40:15.145 the first red flag to
NOTE Confidence: 0.9983724

00:40:15.145 --> 00:40:15.645 identify

NOTE Confidence: 0.9760335
00:40:16.105 --> 00:40:17.625 a Golgi syndrome in a
NOTE Confidence: 0.9760335
00:40:17.625 --> 00:40:18.125 child.
NOTE Confidence: 0.4663086
00:40:19.619 --> 00:40:20.119 Syndrome
NOTE Confidence: 0.844744
00:40:20.500 --> 00:40:22.760 described by, doctor Robert Golin
NOTE Confidence: 0.844744
00:40:22.900 --> 00:40:23.640 in Minneapolis
NOTE Confidence: 0.72021484
00:40:24.579 --> 00:40:24.819 is,
NOTE Confidence: 0.8404541
00:40:25.460 --> 00:40:26.359 a multi,
NOTE Confidence: 0.62060547
00:40:28.020 --> 00:40:28.520 tumoral,
NOTE Confidence: 0.97631836
00:40:29.299 --> 00:40:30.119 cancer predisposition
NOTE Confidence: 0.88256836
00:40:30.500 --> 00:40:31.000 syndrome,
NOTE Confidence: 0.96150714
00:40:31.859 --> 00:40:33.400 with, three genes
NOTE Confidence: 0.92155594
00:40:33.935 --> 00:40:35.535 right now that are associated
NOTE Confidence: 0.92155594
00:40:35.535 --> 00:40:37.614 with, with it. Patch one
NOTE Confidence: 0.92155594
00:40:37.614 --> 00:40:39.775 is the first one. Sufu
NOTE Confidence: 0.92155594
00:40:39.775 --> 00:40:40.815 is another one and then
NOTE Confidence: 0.92155594

00:40:40.815 --> 00:40:41.775 patch two. This is the
NOTE Confidence: 0.92155594

00:40:41.775 --> 00:40:42.975 one that Alan bailed and
NOTE Confidence: 0.92155594

00:40:42.975 --> 00:40:44.655 this is seventeen to ninety
NOTE Confidence: 0.92155594

00:40:44.655 --> 00:40:46.175 percent of cases are associated
NOTE Confidence: 0.92155594

00:40:46.175 --> 00:40:47.395 with patch one abnormalities.
NOTE Confidence: 0.73217773

00:40:48.579 --> 00:40:50.260 We published this page, this
NOTE Confidence: 0.73217773

00:40:50.260 --> 00:40:50.760 case,
NOTE Confidence: 0.75323486

00:40:51.380 --> 00:40:53.300 as a a combination of
NOTE Confidence: 0.75323486

00:40:53.300 --> 00:40:54.280 problem, my sarcoma,
NOTE Confidence: 0.7534668

00:40:54.900 --> 00:40:56.440 and deletion of the patching
NOTE Confidence: 0.55615234

00:40:56.739 --> 00:40:57.400 in syndrome
NOTE Confidence: 0.9245148

00:40:57.940 --> 00:40:59.400 appears in nature oncology,
NOTE Confidence: 0.9970703

00:40:59.940 --> 00:41:01.160 and it has been
NOTE Confidence: 0.89746094

00:41:02.184 --> 00:41:02.684 important,
NOTE Confidence: 0.9845947

00:41:03.224 --> 00:41:04.825 interesting paper in my in
NOTE Confidence: 0.9845947

00:41:04.825 --> 00:41:07.405 in my, personal collection because,

NOTE Confidence: 0.99853516

00:41:08.025 --> 00:41:09.484 it taught me a lot.

NOTE Confidence: 0.99253625

00:41:09.785 --> 00:41:11.145 You can see multiple basal

NOTE Confidence: 0.99253625

00:41:11.145 --> 00:41:11.885 cell carcinomas

NOTE Confidence: 0.8949219

00:41:12.265 --> 00:41:13.545 in these tumors in these

NOTE Confidence: 0.8949219

00:41:13.545 --> 00:41:14.940 patients, and this is how

NOTE Confidence: 0.8949219

00:41:14.940 --> 00:41:16.880 Robert Gordon, Bob Gordon identified

NOTE Confidence: 0.88948566

00:41:17.260 --> 00:41:18.700 the the the syndrome. He

NOTE Confidence: 0.88948566

00:41:18.700 --> 00:41:19.200 was

NOTE Confidence: 0.9944661

00:41:19.500 --> 00:41:20.480 on a bus

NOTE Confidence: 0.9678467

00:41:21.260 --> 00:41:23.260 riding in Minneapolis and saw

NOTE Confidence: 0.9678467

00:41:23.260 --> 00:41:25.020 a patient, and he used

NOTE Confidence: 0.9678467

00:41:25.020 --> 00:41:26.780 to go and say, excuse

NOTE Confidence: 0.9678467

00:41:26.780 --> 00:41:28.155 me. I am doctor Gordon.

NOTE Confidence: 0.9732948

00:41:28.475 --> 00:41:29.435 Do you mind if I

NOTE Confidence: 0.9732948

00:41:29.435 --> 00:41:30.315 speak with you for a

NOTE Confidence: 0.9732948

00:41:30.315 --> 00:41:31.135 for a minute?
NOTE Confidence: 0.95802736

00:41:31.755 --> 00:41:33.035 And they will be, no.
NOTE Confidence: 0.95802736

00:41:33.035 --> 00:41:34.555 No. Sure. And then he
NOTE Confidence: 0.95802736

00:41:34.555 --> 00:41:36.075 will explain that he's concerned
NOTE Confidence: 0.95802736

00:41:36.075 --> 00:41:37.275 with these lesions that the
NOTE Confidence: 0.95802736

00:41:37.275 --> 00:41:38.735 patient has in his face.
NOTE Confidence: 0.90683997

00:41:39.435 --> 00:41:41.515 And then he described the
NOTE Confidence: 0.90683997

00:41:41.515 --> 00:41:42.795 the the the the lesion.
NOTE Confidence: 0.90683997

00:41:42.795 --> 00:41:43.855 This is Bob Gordon.
NOTE Confidence: 0.88183594

00:41:44.730 --> 00:41:46.570 We invited Bob Golin to
NOTE Confidence: 0.88183594

00:41:46.570 --> 00:41:48.250 come to Deepgram rounds here
NOTE Confidence: 0.88183594

00:41:48.250 --> 00:41:49.770 many years ago, a couple
NOTE Confidence: 0.88183594

00:41:49.770 --> 00:41:51.550 years up, before he's dead.
NOTE Confidence: 0.9987305

00:41:52.250 --> 00:41:53.950 And when I invited him,
NOTE Confidence: 0.93828124

00:41:54.969 --> 00:41:56.670 I called his office directly,
NOTE Confidence: 0.9838867

00:41:57.444 --> 00:41:58.664 and, the

NOTE Confidence: 0.9431966
00:41:59.125 --> 00:42:00.265 secretary answered and
NOTE Confidence: 0.9217616
00:42:00.565 --> 00:42:02.325 said, I said, this is
NOTE Confidence: 0.9217616
00:42:02.325 --> 00:42:03.765 doctor Reyes from Yale University.
NOTE Confidence: 0.9217616
00:42:03.765 --> 00:42:04.724 May I speak with doctor
NOTE Confidence: 0.9217616
00:42:04.724 --> 00:42:06.085 Gorlin? I am I am
NOTE Confidence: 0.9217616
00:42:06.085 --> 00:42:08.484 narrating this conversation here, and
NOTE Confidence: 0.9217616
00:42:08.484 --> 00:42:09.844 and she goes, yes. Just
NOTE Confidence: 0.9217616
00:42:09.844 --> 00:42:10.664 a minute, please.
NOTE Confidence: 0.93066406
00:42:11.120 --> 00:42:12.000 And then she comes to
NOTE Confidence: 0.93066406
00:42:12.000 --> 00:42:13.120 the phone and I said,
NOTE Confidence: 0.9133708
00:42:13.520 --> 00:42:15.200 she says, hello? And I
NOTE Confidence: 0.9133708
00:42:15.200 --> 00:42:16.580 said, doctor Golin,
NOTE Confidence: 0.8097331
00:42:17.120 --> 00:42:18.660 si in Spanish.
NOTE Confidence: 0.870474
00:42:19.280 --> 00:42:20.900 Okay? I switched to Spanish
NOTE Confidence: 0.870474
00:42:21.200 --> 00:42:22.880 and I start speaking with
NOTE Confidence: 0.870474

00:42:22.880 --> 00:42:24.340 him and then
NOTE Confidence: 0.93410236
00:42:24.875 --> 00:42:26.315 I switched back to English
NOTE Confidence: 0.93410236
00:42:26.315 --> 00:42:27.515 and I I invited him
NOTE Confidence: 0.93410236
00:42:27.515 --> 00:42:28.075 and all that. And I
NOTE Confidence: 0.93410236
00:42:28.075 --> 00:42:29.515 said, doctor Collin, where did
NOTE Confidence: 0.93410236
00:42:29.515 --> 00:42:31.295 you learn your Spanish?
NOTE Confidence: 0.92578125
00:42:33.674 --> 00:42:34.635 He told me that he
NOTE Confidence: 0.92578125
00:42:34.635 --> 00:42:36.415 was part of a CAA
NOTE Confidence: 0.92578125
00:42:36.714 --> 00:42:37.214 CIA
NOTE Confidence: 0.9951172
00:42:37.515 --> 00:42:38.015 mission
NOTE Confidence: 0.9362793
00:42:39.560 --> 00:42:41.020 in the Nordic countries
NOTE Confidence: 0.84733075
00:42:42.120 --> 00:42:44.040 and that he spent six
NOTE Confidence: 0.84733075
00:42:44.040 --> 00:42:44.540 weeks
NOTE Confidence: 0.99902344
00:42:45.080 --> 00:42:45.580 sharing
NOTE Confidence: 0.99853516
00:42:46.040 --> 00:42:46.780 the room
NOTE Confidence: 0.9523112
00:42:47.080 --> 00:42:48.380 with a Spanish

NOTE Confidence: 0.99902344
00:42:49.000 --> 00:42:49.500 priest
NOTE Confidence: 0.9954834
00:42:50.040 --> 00:42:51.000 that was part of the
NOTE Confidence: 0.9954834
00:42:51.000 --> 00:42:52.460 same CIA mission.
NOTE Confidence: 0.9907924
00:42:53.065 --> 00:42:54.025 I didn't want to know
NOTE Confidence: 0.9907924
00:42:54.025 --> 00:42:54.925 anything else.
NOTE Confidence: 0.9213867
00:42:56.505 --> 00:42:57.785 I didn't bear to ask
NOTE Confidence: 0.9213867
00:42:57.785 --> 00:42:58.685 anything else.
NOTE Confidence: 0.95305526
00:42:59.465 --> 00:43:00.765 She she was a fantastic
NOTE Confidence: 0.95305526
00:43:00.825 --> 00:43:01.965 individual, enormous
NOTE Confidence: 0.89953977
00:43:02.265 --> 00:43:03.625 guy. And when he came
NOTE Confidence: 0.89953977
00:43:03.625 --> 00:43:05.305 into my office, he barely
NOTE Confidence: 0.89953977
00:43:05.305 --> 00:43:07.330 fit. And and he's, told
NOTE Confidence: 0.89953977
00:43:07.330 --> 00:43:08.369 me that he had mantle
NOTE Confidence: 0.89953977
00:43:08.369 --> 00:43:09.190 cell lymphoma.
NOTE Confidence: 0.97365993
00:43:09.489 --> 00:43:11.250 He said, my spleen is
NOTE Confidence: 0.97365993

00:43:11.250 --> 00:43:12.469 the size of Texas.
NOTE Confidence: 0.9809367

00:43:13.090 --> 00:43:14.369 He died a few years
NOTE Confidence: 0.9809367

00:43:14.369 --> 00:43:15.410 later, but he was a
NOTE Confidence: 0.9809367

00:43:15.410 --> 00:43:16.469 fantastic individual.
NOTE Confidence: 0.9260254

00:43:18.369 --> 00:43:19.270 Another example,
NOTE Confidence: 0.9548903

00:43:20.085 --> 00:43:21.605 bloom syndrome. And this is
NOTE Confidence: 0.9548903

00:43:21.605 --> 00:43:23.605 something that I also saw
NOTE Confidence: 0.9548903

00:43:23.605 --> 00:43:24.585 here at Yale.
NOTE Confidence: 0.93029296

00:43:25.364 --> 00:43:27.285 Dan, Pat, Jane was my
NOTE Confidence: 0.93029296

00:43:27.285 --> 00:43:29.045 resident, was a resident at
NOTE Confidence: 0.93029296

00:43:29.045 --> 00:43:30.085 that time. I'm sure that
NOTE Confidence: 0.93029296

00:43:30.085 --> 00:43:31.844 you know who doctor Jane,
NOTE Confidence: 0.93029296

00:43:31.844 --> 00:43:34.025 famous doctor Dampa Jane is.
NOTE Confidence: 0.99658203

00:43:34.325 --> 00:43:35.705 And we saw this
NOTE Confidence: 0.92716473

00:43:38.380 --> 00:43:40.219 patient with a bloom syndrome
NOTE Confidence: 0.92716473

00:43:40.219 --> 00:43:41.600 and a Williams tumor.

NOTE Confidence: 0.96687824
00:43:41.980 --> 00:43:43.580 This is a syndrome that,
NOTE Confidence: 0.96687824
00:43:44.060 --> 00:43:44.560 involves
NOTE Confidence: 0.7949829
00:43:45.100 --> 00:43:47.200 repairing DNA, repairing enzymes.
NOTE Confidence: 0.9915527
00:43:47.820 --> 00:43:49.040 And when they are broken,
NOTE Confidence: 0.9915527
00:43:49.260 --> 00:43:50.719 they don't repair the DNA.
NOTE Confidence: 0.9789063
00:43:51.145 --> 00:43:52.925 This case had an aplasia
NOTE Confidence: 0.8483887
00:43:53.545 --> 00:43:54.765 as you can see here
NOTE Confidence: 0.8483887
00:43:54.985 --> 00:43:55.485 and
NOTE Confidence: 0.99560547
00:43:56.105 --> 00:43:56.925 the sister
NOTE Confidence: 0.9565633
00:43:57.305 --> 00:43:59.385 of this patient developed a
NOTE Confidence: 0.9565633
00:43:59.385 --> 00:43:59.885 hepatocellular
NOTE Confidence: 0.9601237
00:44:00.345 --> 00:44:00.845 carcinoma.
NOTE Confidence: 0.9760742
00:44:01.785 --> 00:44:04.045 Both had the same genetic
NOTE Confidence: 0.9760742
00:44:04.105 --> 00:44:04.605 abnormalities.
NOTE Confidence: 0.9971924
00:44:05.530 --> 00:44:06.810 Two children in the same
NOTE Confidence: 0.9971924

00:44:06.810 --> 00:44:08.270 family with two
NOTE Confidence: 1

00:44:08.650 --> 00:44:09.150 relatively
NOTE Confidence: 0.89782715

00:44:09.450 --> 00:44:10.510 infrequent lesions.
NOTE Confidence: 0.99869794

00:44:11.130 --> 00:44:12.190 They need to
NOTE Confidence: 0.94292533

00:44:12.650 --> 00:44:14.170 rise the red flag for
NOTE Confidence: 0.94292533

00:44:14.170 --> 00:44:15.390 you to pursue it.
NOTE Confidence: 0.98934656

00:44:15.690 --> 00:44:17.290 The pathologist cannot be just
NOTE Confidence: 0.98934656

00:44:17.290 --> 00:44:18.410 sitting in the office and
NOTE Confidence: 0.98934656

00:44:18.410 --> 00:44:18.910 making
NOTE Confidence: 0.9633789

00:44:19.395 --> 00:44:21.075 as my wife says, when
NOTE Confidence: 0.9633789

00:44:21.075 --> 00:44:22.435 when we are working together,
NOTE Confidence: 0.9633789

00:44:22.435 --> 00:44:23.635 she says, oh, you are
NOTE Confidence: 0.9633789

00:44:23.635 --> 00:44:24.915 just giving names to these
NOTE Confidence: 0.9633789

00:44:24.915 --> 00:44:26.275 animals, right, to these,
NOTE Confidence: 0.9815674

00:44:27.155 --> 00:44:29.155 cows. Well, no. We we
NOTE Confidence: 0.9815674

00:44:29.155 --> 00:44:30.375 are part of the clinical

NOTE Confidence: 0.9970703
00:44:31.739 --> 00:44:32.239 advisory
NOTE Confidence: 0.9850098
00:44:33.100 --> 00:44:34.960 team for the patient's family.
NOTE Confidence: 0.99975586
00:44:35.260 --> 00:44:36.320 We need to pursue
NOTE Confidence: 0.98530716
00:44:36.620 --> 00:44:38.060 that they follow it because
NOTE Confidence: 0.98530716
00:44:38.060 --> 00:44:39.100 if they if you don't
NOTE Confidence: 0.98530716
00:44:39.100 --> 00:44:40.300 do that, they may or
NOTE Confidence: 0.98530716
00:44:40.300 --> 00:44:41.900 may not realize that it
NOTE Confidence: 0.98530716
00:44:41.900 --> 00:44:42.640 is important.
NOTE Confidence: 0.99438477
00:44:44.225 --> 00:44:45.105 I don't know if I
NOTE Confidence: 0.99438477
00:44:45.105 --> 00:44:45.425 would,
NOTE Confidence: 0.9932373
00:44:45.905 --> 00:44:47.605 repeat the diagnosis of hepatocellular
NOTE Confidence: 0.8431206
00:44:47.905 --> 00:44:49.425 carcinoma on this particular case
NOTE Confidence: 0.8431206
00:44:49.425 --> 00:44:51.585 because hepatocellular carcinoma in children
NOTE Confidence: 0.8431206
00:44:51.585 --> 00:44:53.185 is relatively rare and is
NOTE Confidence: 0.8431206
00:44:53.185 --> 00:44:54.131 in a process of being
NOTE Confidence: 0.8431206

00:44:54.131 --> 00:44:54.298 reclassified as we speak in
NOTE Confidence: 0.8431206

00:44:54.298 --> 00:44:54.465 the sixth edition of the
NOTE Confidence: 0.8431206

00:44:54.465 --> 00:44:55.215 of the
NOTE Confidence: 0.8526286

00:44:59.839 --> 00:45:02.160 WHO Digestive Tumors book. But
NOTE Confidence: 0.8526286

00:45:02.160 --> 00:45:03.940 we published these these patients
NOTE Confidence: 0.8059082

00:45:04.640 --> 00:45:05.780 with bloom syndrome
NOTE Confidence: 0.7290039

00:45:06.160 --> 00:45:06.480 with,
NOTE Confidence: 0.84160155

00:45:07.040 --> 00:45:07.540 heparocellular
NOTE Confidence: 0.944733

00:45:08.000 --> 00:45:09.520 and WIMS tumor in both
NOTE Confidence: 0.944733

00:45:09.520 --> 00:45:10.480 of them in the same
NOTE Confidence: 0.944733

00:45:10.480 --> 00:45:12.484 family. So those are clues
NOTE Confidence: 0.944733

00:45:12.545 --> 00:45:14.305 you see the same family
NOTE Confidence: 0.944733

00:45:14.305 --> 00:45:16.085 having two children with tumors
NOTE Confidence: 0.8036296

00:45:16.385 --> 00:45:17.444 as not frequent.
NOTE Confidence: 0.7104492

00:45:19.424 --> 00:45:20.244 Bloom syndrome
NOTE Confidence: 0.9932338

00:45:20.545 --> 00:45:22.385 looks like this clinically. They

NOTE Confidence: 0.9932338
00:45:22.385 --> 00:45:23.364 have hypersensitivity
NOTE Confidence: 0.99328613
00:45:24.065 --> 00:45:24.724 to the,
NOTE Confidence: 0.9623128
00:45:25.340 --> 00:45:27.260 sunlight, and they have multiple
NOTE Confidence: 0.9623128
00:45:27.260 --> 00:45:29.520 abnormal abnormalities in the karyotypes
NOTE Confidence: 0.98883057
00:45:29.900 --> 00:45:31.680 due to the abnormal mutations
NOTE Confidence: 0.98883057
00:45:31.820 --> 00:45:32.719 in the repair
NOTE Confidence: 0.93218315
00:45:33.020 --> 00:45:34.700 enzymes of DNA. I'm not
NOTE Confidence: 0.93218315
00:45:34.700 --> 00:45:36.719 gonna detail all these because
NOTE Confidence: 0.93218315
00:45:36.940 --> 00:45:38.300 that's we are gonna run
NOTE Confidence: 0.93218315
00:45:38.300 --> 00:45:39.120 out of time.
NOTE Confidence: 0.9876114
00:45:40.255 --> 00:45:42.415 I'm gonna probably finish with,
NOTE Confidence: 0.9876114
00:45:42.735 --> 00:45:44.575 this group of disorders, multiple
NOTE Confidence: 0.9876114
00:45:44.575 --> 00:45:46.195 endocrine neoplasia syndromes.
NOTE Confidence: 0.97957766
00:45:46.655 --> 00:45:47.935 These are typically seen in
NOTE Confidence: 0.97957766
00:45:47.935 --> 00:45:49.535 adults. We all are familiar
NOTE Confidence: 0.97957766

00:45:49.535 --> 00:45:50.655 with them, but you can
NOTE Confidence: 0.97957766

00:45:50.655 --> 00:45:52.035 also see them in children.
NOTE Confidence: 0.9003906

00:45:54.020 --> 00:45:55.560 This individual here
NOTE Confidence: 0.789917

00:45:56.180 --> 00:45:58.040 is doctor Ivan Carney.
NOTE Confidence: 0.95495605

00:45:58.900 --> 00:46:00.180 I'm sure that many of
NOTE Confidence: 0.95495605

00:46:00.180 --> 00:46:01.940 you have heard of the
NOTE Confidence: 0.95495605

00:46:01.940 --> 00:46:03.000 Kearney syndrome,
NOTE Confidence: 0.9550781

00:46:03.619 --> 00:46:04.840 Kearney's triad,
NOTE Confidence: 0.9268799

00:46:05.235 --> 00:46:07.495 etcetera, etcetera. Kearney's complex.
NOTE Confidence: 0.9561523

00:46:07.875 --> 00:46:09.335 He described all of those.
NOTE Confidence: 0.97281903

00:46:10.115 --> 00:46:11.235 He is sitting with me
NOTE Confidence: 0.97281903

00:46:11.235 --> 00:46:12.435 in my office here at
NOTE Confidence: 0.97281903

00:46:12.435 --> 00:46:14.375 Yale about twenty years ago.
NOTE Confidence: 0.95113117

00:46:14.675 --> 00:46:16.515 I invited him twice, once
NOTE Confidence: 0.95113117

00:46:16.515 --> 00:46:18.115 here and once in Pittsburgh.
NOTE Confidence: 0.95113117

00:46:18.115 --> 00:46:19.575 He was a fantastic guy.

NOTE Confidence: 0.86816406
00:46:19.955 --> 00:46:20.455 Small,
NOTE Confidence: 1
00:46:21.320 --> 00:46:21.820 Irish
NOTE Confidence: 0.8586426
00:46:22.120 --> 00:46:22.620 gentleman,
NOTE Confidence: 0.9555664
00:46:23.160 --> 00:46:24.620 funny, and very,
NOTE Confidence: 0.99869794
00:46:25.080 --> 00:46:26.380 you know, combative.
NOTE Confidence: 0.99853516
00:46:26.920 --> 00:46:27.660 I remember
NOTE Confidence: 0.8959961
00:46:28.520 --> 00:46:29.640 I I remember that,
NOTE Confidence: 0.93644714
00:46:30.360 --> 00:46:32.280 I think Stuart Flynn, told
NOTE Confidence: 0.93644714
00:46:32.280 --> 00:46:33.560 me that he was presenting
NOTE Confidence: 0.93644714
00:46:33.560 --> 00:46:35.320 something, and then Alan Carney
NOTE Confidence: 0.93644714
00:46:35.320 --> 00:46:36.844 was jumping in the audience
NOTE Confidence: 0.93644714
00:46:36.905 --> 00:46:38.505 trying to contradict him and
NOTE Confidence: 0.93644714
00:46:38.505 --> 00:46:39.864 he found him, a very
NOTE Confidence: 0.93644714
00:46:39.864 --> 00:46:41.005 combative spirit.
NOTE Confidence: 0.9267273
00:46:41.945 --> 00:46:43.465 Very nice guy. And,
NOTE Confidence: 0.85932416

00:46:44.665 --> 00:46:45.864 we are referring to the
NOTE Confidence: 0.85932416

00:46:45.864 --> 00:46:47.305 Cipoll Williams Pierce,
NOTE Confidence: 0.94018555

00:46:47.625 --> 00:46:48.605 triad of,
NOTE Confidence: 0.9743071

00:46:48.950 --> 00:46:50.809 individuals that describe the multiple
NOTE Confidence: 0.9743071

00:46:50.869 --> 00:46:51.849 endocrine neoplasia
NOTE Confidence: 0.80194765

00:46:52.150 --> 00:46:53.930 syndromes where had
NOTE Confidence: 0.99820966

00:46:54.390 --> 00:46:55.450 a major role.
NOTE Confidence: 0.9866265

00:46:55.829 --> 00:46:57.029 There are many types of
NOTE Confidence: 0.9866265

00:46:57.029 --> 00:46:58.650 these syndromes, cancer predisposition
NOTE Confidence: 0.88500977

00:46:58.950 --> 00:46:59.450 syndromes.
NOTE Confidence: 0.9654541

00:46:59.989 --> 00:47:01.690 And what happened with,
NOTE Confidence: 0.9817871

00:47:02.915 --> 00:47:04.295 in in in this particular,
NOTE Confidence: 0.9958225

00:47:04.675 --> 00:47:06.035 group of disorders is that
NOTE Confidence: 0.9958225

00:47:06.035 --> 00:47:07.734 we saw two sisters
NOTE Confidence: 0.98046875

00:47:08.515 --> 00:47:09.575 many years ago
NOTE Confidence: 0.99316406

00:47:10.035 --> 00:47:10.935 that came

NOTE Confidence: 0.9995117
00:47:11.714 --> 00:47:12.614 at five
NOTE Confidence: 0.9328342
00:47:13.234 --> 00:47:15.315 years, eleven months, and three
NOTE Confidence: 0.9328342
00:47:15.315 --> 00:47:17.015 years and eight months,
NOTE Confidence: 0.99720985
00:47:17.760 --> 00:47:20.320 Both to be reviewed by
NOTE Confidence: 0.99720985
00:47:20.320 --> 00:47:21.060 the clinicians
NOTE Confidence: 0.9921875
00:47:21.520 --> 00:47:22.640 that were looking at the
NOTE Confidence: 0.9921875
00:47:22.640 --> 00:47:24.500 thyroid because they had
NOTE Confidence: 0.9921875
00:47:24.800 --> 00:47:25.300 a
NOTE Confidence: 0.9831055
00:47:25.760 --> 00:47:27.860 family history of thyroid carcinoma.
NOTE Confidence: 0.9212123
00:47:28.640 --> 00:47:30.239 And I happen to receive
NOTE Confidence: 0.9212123
00:47:30.239 --> 00:47:30.900 the biopsies.
NOTE Confidence: 0.9323543
00:47:31.455 --> 00:47:32.895 We put them together. We
NOTE Confidence: 0.9323543
00:47:32.895 --> 00:47:34.094 we still publish in black
NOTE Confidence: 0.9323543
00:47:34.094 --> 00:47:34.995 and white in
NOTE Confidence: 0.7461751
00:47:35.295 --> 00:47:36.275 those those years.
NOTE Confidence: 0.9920247

00:47:36.575 --> 00:47:37.955 And this individual
NOTE Confidence: 0.95670575

00:47:39.055 --> 00:47:41.055 is relatively well known, Jim
NOTE Confidence: 0.95670575

00:47:41.055 --> 00:47:41.555 Gill,
NOTE Confidence: 0.979047

00:47:42.094 --> 00:47:43.535 whose wife is sitting right
NOTE Confidence: 0.979047

00:47:43.535 --> 00:47:46.140 here. She was my resident
NOTE Confidence: 0.979047

00:47:46.280 --> 00:47:48.040 also and we put together
NOTE Confidence: 0.979047

00:47:48.040 --> 00:47:48.780 this paper
NOTE Confidence: 0.9082031

00:47:49.320 --> 00:47:50.300 and we advise
NOTE Confidence: 0.99902344

00:47:50.840 --> 00:47:51.580 to perform
NOTE Confidence: 0.99780273

00:47:52.040 --> 00:47:52.540 prophylactic
NOTE Confidence: 0.93774414

00:47:53.160 --> 00:47:53.660 thyroidectomy
NOTE Confidence: 0.93434495

00:47:54.520 --> 00:47:56.920 in young children. And, look,
NOTE Confidence: 0.93434495

00:47:56.920 --> 00:47:58.200 this this is an advice
NOTE Confidence: 0.93434495

00:47:58.200 --> 00:47:59.020 that is very
NOTE Confidence: 0.9288424

00:47:59.955 --> 00:48:01.315 delicate to me because you
NOTE Confidence: 0.9288424

00:48:01.315 --> 00:48:02.355 go to the parents and

NOTE Confidence: 0.9288424
00:48:02.355 --> 00:48:03.815 you say, look,
NOTE Confidence: 0.8885091
00:48:04.755 --> 00:48:05.815 you have to
NOTE Confidence: 0.8705706
00:48:06.195 --> 00:48:07.475 two daughters. One of them
NOTE Confidence: 0.8705706
00:48:07.475 --> 00:48:08.375 has unfortunately
NOTE Confidence: 0.9238281
00:48:08.675 --> 00:48:09.815 medullary carcinoma,
NOTE Confidence: 0.989624
00:48:10.275 --> 00:48:11.555 but the other one has
NOTE Confidence: 0.989624
00:48:11.555 --> 00:48:12.775 this change that
NOTE Confidence: 0.95768225
00:48:14.150 --> 00:48:15.590 seems is going to progress
NOTE Confidence: 0.95768225
00:48:15.590 --> 00:48:17.270 to medullary carcinoma. So, you
NOTE Confidence: 0.95768225
00:48:17.270 --> 00:48:18.710 need to recognize that and
NOTE Confidence: 0.95768225
00:48:18.710 --> 00:48:20.469 be part of that exercise
NOTE Confidence: 0.95768225
00:48:20.469 --> 00:48:22.010 in discussing with the patients.
NOTE Confidence: 0.95768225
00:48:22.070 --> 00:48:23.510 We frequently speak with patients.
NOTE Confidence: 0.95768225
00:48:23.510 --> 00:48:24.790 Claudia is a pediatrician, so
NOTE Confidence: 0.95768225
00:48:24.790 --> 00:48:25.750 she she doesn't have a
NOTE Confidence: 0.95768225

00:48:25.750 --> 00:48:26.630 problem with that, but I
NOTE Confidence: 0.95768225

00:48:26.630 --> 00:48:27.530 am not a clinician,
NOTE Confidence: 0.97414726

00:48:27.945 --> 00:48:28.985 But I love to speak
NOTE Confidence: 0.97414726

00:48:28.985 --> 00:48:30.344 with the patients and advise
NOTE Confidence: 0.97414726

00:48:30.344 --> 00:48:31.864 them in conjunction with our
NOTE Confidence: 0.97414726

00:48:31.864 --> 00:48:34.585 clinical colleagues in pediatric oncology.
NOTE Confidence: 0.97414726

00:48:34.585 --> 00:48:36.344 So we published this tale
NOTE Confidence: 0.97414726

00:48:36.344 --> 00:48:37.165 of two sisters
NOTE Confidence: 0.9812012

00:48:37.785 --> 00:48:38.265 with,
NOTE Confidence: 0.9782715

00:48:38.745 --> 00:48:39.565 Jim Gill
NOTE Confidence: 0.8975379

00:48:39.880 --> 00:48:41.400 and doctor Miron Janelle, who
NOTE Confidence: 0.8975379

00:48:41.400 --> 00:48:42.780 was a pediatric endocrinologist
NOTE Confidence: 0.9229126

00:48:43.400 --> 00:48:45.080 at that time directing the
NOTE Confidence: 0.9229126

00:48:45.080 --> 00:48:47.080 program of thyroid cancer in
NOTE Confidence: 0.9229126

00:48:47.080 --> 00:48:47.980 Yale University,
NOTE Confidence: 0.9970703

00:48:48.359 --> 00:48:49.660 and it was a fantastic

NOTE Confidence: 0.9885254

00:48:49.960 --> 00:48:50.460 experience.

NOTE Confidence: 0.92979944

00:48:52.224 --> 00:48:54.065 Multiple endocrine neoplasia has many

NOTE Confidence: 0.92979944

00:48:54.065 --> 00:48:55.765 different expressions, histologically

NOTE Confidence: 0.96261394

00:48:56.145 --> 00:48:58.645 speaking. It has multiple neurofibromas,

NOTE Confidence: 0.9287342

00:48:59.185 --> 00:49:01.185 neuromas, etcetera and you have

NOTE Confidence: 0.9287342

00:49:01.185 --> 00:49:01.765 to be

NOTE Confidence: 0.99576825

00:49:02.145 --> 00:49:03.125 able to recognize

NOTE Confidence: 0.9813802

00:49:03.505 --> 00:49:05.344 the danger that exists when

NOTE Confidence: 0.9813802

00:49:05.344 --> 00:49:06.864 you see lesions like this

NOTE Confidence: 0.9813802

00:49:06.864 --> 00:49:08.900 in young children. Young children

NOTE Confidence: 0.9813802

00:49:09.040 --> 00:49:10.719 should not get sick and

NOTE Confidence: 0.9813802

00:49:10.719 --> 00:49:12.500 if they do, they usually

NOTE Confidence: 0.9813802

00:49:12.559 --> 00:49:14.420 get well with no help

NOTE Confidence: 0.9532508

00:49:14.880 --> 00:49:16.400 but when they don't, you

NOTE Confidence: 0.9532508

00:49:16.400 --> 00:49:18.160 need to pay attention to

NOTE Confidence: 0.9532508

00:49:18.160 --> 00:49:18.660 them.
NOTE Confidence: 0.98494464

00:49:19.599 --> 00:49:20.864 I had a few more
NOTE Confidence: 0.98494464

00:49:20.864 --> 00:49:22.145 examples that I am not
NOTE Confidence: 0.98494464

00:49:22.145 --> 00:49:23.585 going to detail because I'm
NOTE Confidence: 0.98494464

00:49:23.585 --> 00:49:24.465 not going to,
NOTE Confidence: 0.876709

00:49:24.945 --> 00:49:26.165 spend, well, maybe,
NOTE Confidence: 0.99833983

00:49:26.625 --> 00:49:27.665 just to show you that
NOTE Confidence: 0.99833983

00:49:27.665 --> 00:49:29.364 there are several other associations
NOTE Confidence: 0.9151166

00:49:29.985 --> 00:49:31.745 of involving the genital syndrome.
NOTE Confidence: 0.9151166

00:49:31.745 --> 00:49:33.685 For example, the genital area,
NOTE Confidence: 0.9151166

00:49:33.800 --> 00:49:35.960 for example, Frasier syndrome where
NOTE Confidence: 0.9151166

00:49:35.960 --> 00:49:37.320 you can have mutations in
NOTE Confidence: 0.9151166

00:49:37.320 --> 00:49:37.820 WT
NOTE Confidence: 0.45056152

00:49:38.280 --> 00:49:38.760 one,
NOTE Confidence: 0.956448

00:49:39.080 --> 00:49:40.920 gene with different types of
NOTE Confidence: 0.956448

00:49:40.920 --> 00:49:42.920 mutation that can lead to

NOTE Confidence: 0.956448
00:49:42.920 --> 00:49:43.900 different syndromes
NOTE Confidence: 0.961084
00:49:44.359 --> 00:49:45.880 depending on the codon that
NOTE Confidence: 0.961084
00:49:45.880 --> 00:49:47.320 is involved. And you can
NOTE Confidence: 0.961084
00:49:47.320 --> 00:49:49.420 have abnormalities in the kidney
NOTE Confidence: 0.9774292
00:49:50.094 --> 00:49:51.215 and in the gonads. You
NOTE Confidence: 0.9774292
00:49:51.215 --> 00:49:52.275 can have gonadoglastoma
NOTE Confidence: 0.94737816
00:49:53.135 --> 00:49:54.735 like this. When you see
NOTE Confidence: 0.94737816
00:49:54.735 --> 00:49:56.495 different the differences in sex
NOTE Confidence: 0.94737816
00:49:56.495 --> 00:49:58.175 development like a strict gonad
NOTE Confidence: 0.94737816
00:49:58.175 --> 00:49:59.795 that you see here associated
NOTE Confidence: 0.94737816
00:49:59.855 --> 00:50:00.755 with a gonadoglastoma
NOTE Confidence: 0.9852295
00:50:01.855 --> 00:50:03.535 and then you pursue the
NOTE Confidence: 0.9852295
00:50:03.535 --> 00:50:05.075 genetics, you can
NOTE Confidence: 0.98791504
00:50:05.670 --> 00:50:07.130 diagnose a cancer predisposition
NOTE Confidence: 1
00:50:07.510 --> 00:50:08.010 syndrome.
NOTE Confidence: 1

00:50:09.349 --> 00:50:09.849 So
NOTE Confidence: 0.936246
00:50:10.469 --> 00:50:12.550 the major non CNS pediatric
NOTE Confidence: 0.936246
00:50:12.550 --> 00:50:15.210 tumor categories are, mentioned here.
NOTE Confidence: 0.936246
00:50:15.349 --> 00:50:16.469 Most of them I have
NOTE Confidence: 0.936246
00:50:16.469 --> 00:50:17.989 touched on, but I want
NOTE Confidence: 0.936246
00:50:17.989 --> 00:50:18.550 you to,
NOTE Confidence: 0.99401855
00:50:19.430 --> 00:50:20.809 take home these messages.
NOTE Confidence: 0.9942491
00:50:22.125 --> 00:50:23.965 Ten percent of pediatric solid
NOTE Confidence: 0.9942491
00:50:23.965 --> 00:50:25.585 tumors represent cancer predisposition
NOTE Confidence: 0.9995117
00:50:25.965 --> 00:50:26.465 syndromes.
NOTE Confidence: 0.9998373
00:50:26.925 --> 00:50:27.665 The histopathology
NOTE Confidence: 0.9930827
00:50:28.364 --> 00:50:30.305 frequently gives you the sentinel
NOTE Confidence: 0.9930827
00:50:30.364 --> 00:50:30.864 clue
NOTE Confidence: 0.9876302
00:50:31.165 --> 00:50:32.605 to start working on those
NOTE Confidence: 0.9876302
00:50:32.605 --> 00:50:33.105 patients.
NOTE Confidence: 0.9954529
00:50:33.965 --> 00:50:35.405 You have to correlate genetics

NOTE Confidence: 0.9954529
00:50:35.405 --> 00:50:36.625 with clinical data.
NOTE Confidence: 0.9538574
00:50:37.510 --> 00:50:38.330 You the pathologist
NOTE Confidence: 0.99609375
00:50:38.790 --> 00:50:39.290 identifies
NOTE Confidence: 0.94195557
00:50:39.590 --> 00:50:41.210 patients at risk frequently.
NOTE Confidence: 0.9969308
00:50:41.989 --> 00:50:43.270 As long as you think
NOTE Confidence: 0.9969308
00:50:43.270 --> 00:50:43.930 of this,
NOTE Confidence: 0.97706825
00:50:44.310 --> 00:50:45.690 you can just pass it
NOTE Confidence: 0.97706825
00:50:45.830 --> 00:50:47.610 one more slide and next
NOTE Confidence: 0.97706825
00:50:47.750 --> 00:50:48.970 and you miss it.
NOTE Confidence: 0.9980469
00:50:49.350 --> 00:50:50.330 And collaboration
NOTE Confidence: 0.9353841
00:50:50.790 --> 00:50:51.690 with our clinical,
NOTE Confidence: 0.9937134
00:50:52.185 --> 00:50:53.405 genetic, oncological,
NOTE Confidence: 0.94750977
00:50:54.105 --> 00:50:54.605 surgical,
NOTE Confidence: 0.8010254
00:50:55.545 --> 00:50:56.685 pediatric specialists
NOTE Confidence: 0.9802246
00:50:57.225 --> 00:50:58.445 assures adequate
NOTE Confidence: 0.9838867

00:50:59.065 --> 00:50:59.565 precision
NOTE Confidence: 0.99365234

00:50:59.945 --> 00:51:00.445 diagnosis.
NOTE Confidence: 0.9268144

00:51:01.065 --> 00:51:02.344 Okay? So there is a
NOTE Confidence: 0.9268144

00:51:02.344 --> 00:51:03.864 great variety great variety of
NOTE Confidence: 0.9268144

00:51:03.864 --> 00:51:06.185 syndromes predisposing to cancer that
NOTE Confidence: 0.9268144

00:51:06.185 --> 00:51:07.165 appear in childhood,
NOTE Confidence: 0.94558376

00:51:07.530 --> 00:51:08.750 The pediatric pathologist,
NOTE Confidence: 0.97546387

00:51:09.290 --> 00:51:10.750 the first to detect suspicious
NOTE Confidence: 0.97546387

00:51:10.810 --> 00:51:12.250 or diagnostic findings in these
NOTE Confidence: 0.97546387

00:51:12.250 --> 00:51:13.630 syndromes, not infrequently.
NOTE Confidence: 0.9844933

00:51:14.170 --> 00:51:16.250 And the developing organism has
NOTE Confidence: 0.9844933

00:51:16.250 --> 00:51:18.030 highly variable ways of phenotypic
NOTE Confidence: 0.9844933

00:51:18.250 --> 00:51:18.750 expression
NOTE Confidence: 0.95772296

00:51:19.050 --> 00:51:20.510 with the pediatric pathologist,
NOTE Confidence: 0.9391646

00:51:21.370 --> 00:51:23.605 which with the with which
NOTE Confidence: 0.9391646

00:51:23.605 --> 00:51:25.525 the pediatric pathologist should be

NOTE Confidence: 0.9391646
00:51:25.525 --> 00:51:26.025 intimately
NOTE Confidence: 0.9883626
00:51:26.325 --> 00:51:27.785 familiarized. If you don't
NOTE Confidence: 0.96128446
00:51:28.085 --> 00:51:29.925 recognize these lesions, you miss
NOTE Confidence: 0.96128446
00:51:29.925 --> 00:51:31.445 an important diagnosis that will
NOTE Confidence: 0.96128446
00:51:31.445 --> 00:51:33.125 affect not only that patient
NOTE Confidence: 0.96128446
00:51:33.125 --> 00:51:34.345 but the entire family.
NOTE Confidence: 0.95107424
00:51:35.765 --> 00:51:37.225 And I finish with that.
NOTE Confidence: 0.90866697
00:51:39.820 --> 00:51:41.580 You may recognize yourself some
NOTE Confidence: 0.90866697
00:51:41.580 --> 00:51:43.660 of yourselves here. I couldn't
NOTE Confidence: 0.90866697
00:51:43.660 --> 00:51:45.520 put everyone but, you know,
NOTE Confidence: 0.99316406
00:51:46.300 --> 00:51:46.800 nice
NOTE Confidence: 0.77144367
00:51:47.420 --> 00:51:49.099 memories from Jim Gill, Marie
NOTE Confidence: 0.77144367
00:51:49.099 --> 00:51:49.599 Rivera,
NOTE Confidence: 0.82470703
00:51:50.140 --> 00:51:50.380 and,
NOTE Confidence: 0.989043
00:51:51.335 --> 00:51:52.375 well, I am not going
NOTE Confidence: 0.989043

00:51:52.375 --> 00:51:54.055 to name everyone because you
NOTE Confidence: 0.989043

00:51:54.055 --> 00:51:55.815 know who they are. Thank
NOTE Confidence: 0.989043

00:51:55.815 --> 00:51:56.934 you very much for the
NOTE Confidence: 0.989043

00:51:56.934 --> 00:51:58.954 honor of, visiting Yale again.
NOTE Confidence: 0.99853516

00:51:59.654 --> 00:52:00.394 Thank you.
NOTE Confidence: 0.9758301

00:52:04.560 --> 00:52:05.460 Any questions,
NOTE Confidence: 0.9975586

00:52:06.080 --> 00:52:06.580 comments?
NOTE Confidence: 0.9512451

00:52:11.040 --> 00:52:12.160 I I know that you
NOTE Confidence: 0.9512451

00:52:12.160 --> 00:52:13.860 always have a question, Joe.
NOTE Confidence: 0.60057205

00:52:17.075 --> 00:52:17.955 You know what they did?
NOTE Confidence: 0.60057205

00:52:17.955 --> 00:52:19.555 You need sequencing or sequence,
NOTE Confidence: 0.60057205

00:52:19.555 --> 00:52:20.995 whatever, like, all it is
NOTE Confidence: 0.60057205

00:52:20.995 --> 00:52:21.795 is that they need all
NOTE Confidence: 0.60057205

00:52:21.795 --> 00:52:22.535 good secrets.
NOTE Confidence: 0.7659302

00:52:23.395 --> 00:52:24.355 Can you share with us
NOTE Confidence: 0.7659302

00:52:24.355 --> 00:52:24.995 what do you think are

NOTE Confidence: 0.7659302
00:52:24.995 --> 00:52:26.035 driving the depth of just
NOTE Confidence: 0.7659302
00:52:26.035 --> 00:52:27.715 a lot of things we
NOTE Confidence: 0.7659302
00:52:27.715 --> 00:52:28.455 don't recognize
NOTE Confidence: 0.90511066
00:52:28.755 --> 00:52:29.795 that are gonna come out
NOTE Confidence: 0.90511066
00:52:29.795 --> 00:52:30.775 of all that sequence?
NOTE Confidence: 0.99527997
00:52:31.860 --> 00:52:33.380 Yeah. It's a it's a
NOTE Confidence: 0.99527997
00:52:33.380 --> 00:52:33.880 very
NOTE Confidence: 1
00:52:34.340 --> 00:52:34.840 important
NOTE Confidence: 0.9585724
00:52:35.140 --> 00:52:36.420 point, John, that you raised.
NOTE Confidence: 0.9585724
00:52:36.420 --> 00:52:37.780 And, believe me, it's a
NOTE Confidence: 0.9585724
00:52:37.780 --> 00:52:39.640 very hot point of discussion
NOTE Confidence: 0.9585724
00:52:39.700 --> 00:52:40.200 now.
NOTE Confidence: 0.9752308
00:52:40.740 --> 00:52:42.340 And Jose and I exchanged
NOTE Confidence: 0.9752308
00:52:42.340 --> 00:52:43.560 a couple of years ago
NOTE Confidence: 0.9752308
00:52:43.620 --> 00:52:45.635 papers on the relevance of
NOTE Confidence: 0.9752308

00:52:45.635 --> 00:52:47.155 phenotype and genetic changes and
NOTE Confidence: 0.9752308

00:52:47.155 --> 00:52:47.815 the correlation
NOTE Confidence: 0.96272784

00:52:48.195 --> 00:52:49.955 of these two things. And
NOTE Confidence: 0.96272784

00:52:49.955 --> 00:52:51.555 right now, the WHO is
NOTE Confidence: 0.96272784

00:52:51.555 --> 00:52:52.614 suffering from,
NOTE Confidence: 0.9584612

00:52:54.355 --> 00:52:55.795 I wouldn't say attacks, but
NOTE Confidence: 0.9584612

00:52:55.795 --> 00:52:57.555 questioning, severe questioning from our
NOTE Confidence: 0.9584612

00:52:57.555 --> 00:52:59.474 clinical colleagues that say, well,
NOTE Confidence: 0.9584612

00:52:59.474 --> 00:53:01.360 you know, histology doesn't matter
NOTE Confidence: 0.9584612

00:53:01.360 --> 00:53:01.860 anymore.
NOTE Confidence: 0.9791838

00:53:02.320 --> 00:53:03.700 You just need to sequence.
NOTE Confidence: 0.9791838

00:53:03.840 --> 00:53:04.800 You just need to put
NOTE Confidence: 0.9791838

00:53:04.800 --> 00:53:07.200 the patient's tissue through, genetic
NOTE Confidence: 0.9791838

00:53:07.200 --> 00:53:09.060 analysis and identify deletion.
NOTE Confidence: 0.9415394

00:53:09.760 --> 00:53:11.200 And I wish that could
NOTE Confidence: 0.9415394

00:53:11.200 --> 00:53:12.739 be possible but it isn't

NOTE Confidence: 0.9415394
00:53:12.800 --> 00:53:13.300 because
NOTE Confidence: 0.99690753
00:53:15.965 --> 00:53:18.305 many different types of genetic
NOTE Confidence: 0.99690753
00:53:18.445 --> 00:53:18.945 abnormalities
NOTE Confidence: 0.9973958
00:53:19.245 --> 00:53:20.385 that are identifiable
NOTE Confidence: 0.9758301
00:53:20.925 --> 00:53:23.405 by sequencing different modalities of
NOTE Confidence: 0.9758301
00:53:23.405 --> 00:53:23.905 sequencing
NOTE Confidence: 0.81396484
00:53:24.844 --> 00:53:25.344 generate
NOTE Confidence: 0.9998372
00:53:25.885 --> 00:53:27.344 completely different phenotypes
NOTE Confidence: 0.97176105
00:53:27.965 --> 00:53:29.325 depending on a number of
NOTE Confidence: 0.97176105
00:53:29.325 --> 00:53:29.825 other
NOTE Confidence: 0.9187012
00:53:30.450 --> 00:53:30.950 things,
NOTE Confidence: 0.99365234
00:53:31.410 --> 00:53:31.910 epigenetics
NOTE Confidence: 0.81811523
00:53:32.369 --> 00:53:33.270 and environmental
NOTE Confidence: 0.98706055
00:53:33.890 --> 00:53:34.390 circumstances.
NOTE Confidence: 0.89338684
00:53:35.330 --> 00:53:37.170 The phenotype, which is what
NOTE Confidence: 0.89338684

00:53:37.170 --> 00:53:38.630 we pathologists see,
NOTE Confidence: 0.9326579

00:53:39.010 --> 00:53:40.310 macro or microscopically,
NOTE Confidence: 0.9827881

00:53:41.810 --> 00:53:43.430 is really the absolute
NOTE Confidence: 0.9248047

00:53:43.730 --> 00:53:44.230 unquestionable
NOTE Confidence: 0.9440918

00:53:44.770 --> 00:53:46.155 end result of the
NOTE Confidence: 0.9926758

00:53:46.955 --> 00:53:47.855 the genetic expression
NOTE Confidence: 0.99519855

00:53:48.795 --> 00:53:49.855 of an organism.
NOTE Confidence: 0.9976562

00:53:50.635 --> 00:53:52.094 The phenotype of a tissue
NOTE Confidence: 0.92626953

00:53:53.275 --> 00:53:55.855 contains the information to understand
NOTE Confidence: 0.9980469

00:53:56.235 --> 00:53:57.055 how the genetics
NOTE Confidence: 0.9869449

00:53:57.515 --> 00:53:59.275 led to that change, but
NOTE Confidence: 0.9869449

00:53:59.275 --> 00:54:00.735 the reverse is not true.
NOTE Confidence: 0.9869449

00:54:00.980 --> 00:54:02.500 If you only see the
NOTE Confidence: 0.9869449

00:54:02.500 --> 00:54:03.559 part of the genetics,
NOTE Confidence: 0.9995117

00:54:04.099 --> 00:54:05.960 you will miss certain conditions.
NOTE Confidence: 1

00:54:06.660 --> 00:54:07.160 So

NOTE Confidence: 0.9998779
00:54:07.619 --> 00:54:08.739 I hope that we are
NOTE Confidence: 0.9998779
00:54:08.739 --> 00:54:09.640 able to develop
NOTE Confidence: 0.9921875
00:54:10.099 --> 00:54:12.180 additional instruments on both sides
NOTE Confidence: 0.9921875
00:54:12.180 --> 00:54:13.640 of the operation, the genetic
NOTE Confidence: 0.9921875
00:54:13.779 --> 00:54:14.279 analysis
NOTE Confidence: 0.8541667
00:54:14.900 --> 00:54:15.799 and the histological
NOTE Confidence: 0.8695475
00:54:17.195 --> 00:54:19.855 structural, ultra structural, etcetera analysis
NOTE Confidence: 0.87109375
00:54:20.475 --> 00:54:21.535 to bring this
NOTE Confidence: 0.98718905
00:54:21.995 --> 00:54:23.675 to a uniform reality that
NOTE Confidence: 0.98718905
00:54:23.675 --> 00:54:25.594 can serve and guide us
NOTE Confidence: 0.98718905
00:54:25.594 --> 00:54:27.755 for diagnosis and treatment. This
NOTE Confidence: 0.98718905
00:54:27.755 --> 00:54:29.535 issue was pointed at
NOTE Confidence: 0.9642334
00:54:30.280 --> 00:54:31.560 in a recent paper that
NOTE Confidence: 0.9642334
00:54:31.560 --> 00:54:32.840 we, the editorial board of
NOTE Confidence: 0.9642334
00:54:32.840 --> 00:54:33.660 the WHO
NOTE Confidence: 0.7897678

00:54:34.760 --> 00:54:36.440 books published in the Lancet
NOTE Confidence: 0.7897678

00:54:36.440 --> 00:54:36.940 Oncology
NOTE Confidence: 0.93457973

00:54:37.240 --> 00:54:39.000 Journal a few months ago,
NOTE Confidence: 0.93457973

00:54:39.000 --> 00:54:40.760 last year, where we made
NOTE Confidence: 0.93457973

00:54:40.760 --> 00:54:42.200 the case that, yes, we
NOTE Confidence: 0.93457973

00:54:42.200 --> 00:54:44.280 understand that genetic analysis is
NOTE Confidence: 0.93457973

00:54:44.280 --> 00:54:45.560 more and more and more
NOTE Confidence: 0.93457973

00:54:45.560 --> 00:54:46.060 important,
NOTE Confidence: 0.99869794

00:54:46.415 --> 00:54:47.474 but we cannot
NOTE Confidence: 1

00:54:47.935 --> 00:54:48.435 eliminate
NOTE Confidence: 0.99088544

00:54:49.135 --> 00:54:50.575 the the role of the
NOTE Confidence: 0.99088544

00:54:50.575 --> 00:54:51.075 histological
NOTE Confidence: 0.964224

00:54:51.375 --> 00:54:53.135 analysis because we miss the
NOTE Confidence: 0.964224

00:54:53.135 --> 00:54:55.395 phenotype changes, and we miss
NOTE Confidence: 0.964224

00:54:55.535 --> 00:54:57.155 the precision diagnosis.
NOTE Confidence: 0.9818685

00:54:57.535 --> 00:54:58.869 So I don't know if

NOTE Confidence: 0.9818685
00:54:58.869 --> 00:55:00.790 that is a very wishy
NOTE Confidence: 0.9818685
00:55:00.790 --> 00:55:02.550 washy response to you, but
NOTE Confidence: 0.98413086
00:55:05.750 --> 00:55:06.650 We we
NOTE Confidence: 0.80489093
00:55:07.750 --> 00:55:08.950 the whole career has been
NOTE Confidence: 0.80489093
00:55:08.950 --> 00:55:09.450 identified
NOTE Confidence: 0.907959
00:55:09.750 --> 00:55:10.570 new lesions.
NOTE Confidence: 0.9433594
00:55:11.424 --> 00:55:12.625 Now you have a huge
NOTE Confidence: 0.9433594
00:55:12.625 --> 00:55:14.325 resource in the genomic database,
NOTE Confidence: 0.94329834
00:55:15.344 --> 00:55:16.864 and so it should be
NOTE Confidence: 0.94329834
00:55:16.864 --> 00:55:18.005 a pointer that
NOTE Confidence: 0.88479614
00:55:18.305 --> 00:55:19.105 better go back and look
NOTE Confidence: 0.88479614
00:55:19.105 --> 00:55:20.385 at these stations because there's
NOTE Confidence: 0.88479614
00:55:20.385 --> 00:55:21.904 probably something that we hadn't
NOTE Confidence: 0.88479614
00:55:21.904 --> 00:55:22.404 appreciated.
NOTE Confidence: 0.77955043
00:55:23.570 --> 00:55:24.630 Do they set the enemy's
NOTE Confidence: 0.77955043

00:55:24.690 --> 00:55:26.089 valuable? Because it's so fast

NOTE Confidence: 0.77955043

00:55:26.089 --> 00:55:27.590 and effective. It is right.

NOTE Confidence: 0.77955043

00:55:27.730 --> 00:55:29.810 Absolutely. And I would love

NOTE Confidence: 0.77955043

00:55:29.810 --> 00:55:30.850 to be in a project

NOTE Confidence: 0.77955043

00:55:30.850 --> 00:55:31.350 where

NOTE Confidence: 0.96916395

00:55:31.890 --> 00:55:33.090 we look at the genetic

NOTE Confidence: 0.96916395

00:55:33.090 --> 00:55:34.210 changes, and then we go

NOTE Confidence: 0.96916395

00:55:34.210 --> 00:55:35.250 back to the tissues and

NOTE Confidence: 0.96916395

00:55:35.250 --> 00:55:36.610 say, okay. How how did

NOTE Confidence: 0.96916395

00:55:36.610 --> 00:55:37.910 this manifest histologically?

NOTE Confidence: 0.9835693

00:55:38.735 --> 00:55:40.175 That is being done in

NOTE Confidence: 0.9835693

00:55:40.175 --> 00:55:41.715 some places, but it's still

NOTE Confidence: 0.9199219

00:55:42.495 --> 00:55:44.355 early to say. Mina.

NOTE Confidence: 0.8901367

00:55:49.215 --> 00:55:49.715 Mina.

NOTE Confidence: 0.9433245

00:55:59.039 --> 00:56:00.239 I'm sorry. I couldn't hear

NOTE Confidence: 0.9433245

00:56:00.239 --> 00:56:00.819 you. Why?

NOTE Confidence: 0.60375977
00:56:04.319 --> 00:56:04.819 Oh.
NOTE Confidence: 0.99543643
00:56:07.175 --> 00:56:08.614 That's an interesting point. I
NOTE Confidence: 0.99543643
00:56:08.614 --> 00:56:09.915 don't know if we know
NOTE Confidence: 0.99543643
00:56:09.975 --> 00:56:11.675 exactly how the,
NOTE Confidence: 0.79299927
00:56:12.135 --> 00:56:13.435 effect of the one
NOTE Confidence: 0.9863281
00:56:13.815 --> 00:56:14.315 mutation
NOTE Confidence: 0.95118433
00:56:14.614 --> 00:56:16.555 leads to a cystic degeneration.
NOTE Confidence: 0.95118433
00:56:16.695 --> 00:56:17.735 I think that this well,
NOTE Confidence: 0.95118433
00:56:17.735 --> 00:56:18.795 the cystic formation.
NOTE Confidence: 0.99822444
00:56:19.219 --> 00:56:20.660 I think that the cystic
NOTE Confidence: 0.99822444
00:56:20.660 --> 00:56:22.500 formation may have more to
NOTE Confidence: 0.99822444
00:56:22.500 --> 00:56:23.000 do
NOTE Confidence: 0.992041
00:56:23.380 --> 00:56:25.559 with the specific environment histologically
NOTE Confidence: 0.9253743
00:56:25.940 --> 00:56:26.839 that it happens.
NOTE Confidence: 0.9718506
00:56:27.219 --> 00:56:28.420 In the lung, for example,
NOTE Confidence: 0.9718506

00:56:28.420 --> 00:56:30.359 is relatively easy to understand

NOTE Confidence: 0.9718506

00:56:30.420 --> 00:56:31.859 that there there is a

NOTE Confidence: 0.9718506

00:56:31.859 --> 00:56:33.480 flow of air around, etcetera.

NOTE Confidence: 0.95550746

00:56:33.855 --> 00:56:34.734 But I don't know in

NOTE Confidence: 0.95550746

00:56:34.734 --> 00:56:36.015 other places that are solid

NOTE Confidence: 0.95550746

00:56:36.015 --> 00:56:37.855 organs not exposed to these

NOTE Confidence: 0.95550746

00:56:37.855 --> 00:56:39.455 types of pressures. I really

NOTE Confidence: 0.95550746

00:56:39.455 --> 00:56:40.655 don't know. I don't know

NOTE Confidence: 0.95550746

00:56:40.655 --> 00:56:42.355 if anybody knows if,

NOTE Confidence: 0.89038086

00:57:14.075 --> 00:57:15.355 In in the kidney too.

NOTE Confidence: 0.89038086

00:57:15.355 --> 00:57:16.895 In the kidney, cystic nephroma

NOTE Confidence: 0.94767255

00:57:17.460 --> 00:57:19.400 is, by definition, cystic.

NOTE Confidence: 0.62895787

00:57:19.780 --> 00:57:21.300 Yes. I think of the

NOTE Confidence: 0.62895787

00:57:21.300 --> 00:57:22.339 design. Yes. Yes. Yes. But

NOTE Confidence: 0.62895787

00:57:22.339 --> 00:57:23.533 the on the

NOTE Confidence: 0.82910156

00:57:24.660 --> 00:57:25.160 Yeah.

NOTE Confidence: 0.9358724
00:57:29.140 --> 00:57:30.580 Jose? To go to go
NOTE Confidence: 0.9358724
00:57:30.580 --> 00:57:31.640 back to the sequence,
NOTE Confidence: 0.54577637
00:57:45.265 --> 00:57:45.765 Right.
NOTE Confidence: 0.90234375
00:57:47.579 --> 00:57:49.039 And it is clear that
NOTE Confidence: 0.9223633
00:57:49.339 --> 00:57:49.839 epigenetics
NOTE Confidence: 0.6623596
00:57:51.980 --> 00:57:53.420 is one of the major
NOTE Confidence: 0.6623596
00:57:53.420 --> 00:57:54.880 piece when we're sent to
NOTE Confidence: 0.7733154
00:57:55.660 --> 00:57:56.480 that. That means
NOTE Confidence: 0.65818954
00:57:56.940 --> 00:57:58.319 cancer, human enhancers,
NOTE Confidence: 0.9666748
00:57:59.259 --> 00:57:59.920 the whole.
NOTE Confidence: 0.87597656
00:58:16.290 --> 00:58:16.790 Yep.
NOTE Confidence: 0.24902344
00:58:17.570 --> 00:58:18.070 I
NOTE Confidence: 0.9321289
00:58:20.930 --> 00:58:22.210 think that your comment is
NOTE Confidence: 0.9321289
00:58:22.210 --> 00:58:22.710 perfectly
NOTE Confidence: 0.725708
00:58:23.090 --> 00:58:23.590 correct.
NOTE Confidence: 0.9574585

00:58:24.130 --> 00:58:25.890 And we are pushing for
NOTE Confidence: 0.9574585

00:58:25.970 --> 00:58:27.750 we as humans are pushing
NOTE Confidence: 0.9574585

00:58:27.810 --> 00:58:28.550 for that.
NOTE Confidence: 0.99853516

00:58:28.850 --> 00:58:29.350 However,
NOTE Confidence: 0.9980469

00:58:30.130 --> 00:58:31.110 it has had
NOTE Confidence: 0.9847819

00:58:32.325 --> 00:58:34.405 some undesirable effects in the
NOTE Confidence: 0.9847819

00:58:34.405 --> 00:58:34.905 community.
NOTE Confidence: 0.98652345

00:58:35.525 --> 00:58:36.965 One of the examples that
NOTE Confidence: 0.98652345

00:58:36.965 --> 00:58:38.165 I was discussing this morning
NOTE Confidence: 0.98652345

00:58:38.165 --> 00:58:40.025 with Anita Hudner was that
NOTE Confidence: 0.8741048

00:58:40.325 --> 00:58:42.165 the p the CNS five
NOTE Confidence: 0.8741048

00:58:42.165 --> 00:58:43.385 edition of the WHO,
NOTE Confidence: 0.97929686

00:58:43.900 --> 00:58:45.580 the central nervous system tumor
NOTE Confidence: 0.97929686

00:58:45.580 --> 00:58:47.360 volume of the WHO incorporates
NOTE Confidence: 0.9975586

00:58:47.980 --> 00:58:50.320 a massive amount of sequencing
NOTE Confidence: 0.9975586

00:58:50.540 --> 00:58:51.040 and

NOTE Confidence: 0.9552409
00:58:51.420 --> 00:58:53.040 methylation, which is epigenetic
NOTE Confidence: 0.9980469
00:58:53.340 --> 00:58:53.840 changes
NOTE Confidence: 0.5168457
00:58:54.220 --> 00:58:54.720 analysis.
NOTE Confidence: 0.9941406
00:58:55.020 --> 00:58:55.920 And it incorporates
NOTE Confidence: 0.9411926
00:58:56.220 --> 00:58:57.840 them, many of them as
NOTE Confidence: 0.9411926
00:58:57.900 --> 00:58:59.440 essential rather than
NOTE Confidence: 0.9844021
00:58:59.775 --> 00:59:01.694 desirable criteria. And the problem
NOTE Confidence: 0.9844021
00:59:01.694 --> 00:59:03.315 with that is that
NOTE Confidence: 0.98616534
00:59:03.694 --> 00:59:05.135 the over ninety percent of
NOTE Confidence: 0.98616534
00:59:05.135 --> 00:59:06.575 the population in the world
NOTE Confidence: 0.98616534
00:59:06.575 --> 00:59:08.115 has no access to that.
NOTE Confidence: 0.98799956
00:59:08.575 --> 00:59:10.674 So, yes, we need to
NOTE Confidence: 0.98799956
00:59:10.815 --> 00:59:12.910 be able to develop those
NOTE Confidence: 0.98799956
00:59:13.130 --> 00:59:14.990 approaches, those techniques
NOTE Confidence: 0.9859619
00:59:15.690 --> 00:59:17.630 to identify these changes
NOTE Confidence: 0.96435547

00:59:17.930 --> 00:59:20.010 but we cannot demand that
NOTE Confidence: 0.96435547

00:59:20.010 --> 00:59:20.510 everyone
NOTE Confidence: 0.9995117

00:59:21.130 --> 00:59:22.090 has to be able to
NOTE Confidence: 0.9995117

00:59:22.090 --> 00:59:22.590 diagnose
NOTE Confidence: 0.9588068

00:59:23.210 --> 00:59:25.155 a tumor with not only
NOTE Confidence: 0.9588068

00:59:25.155 --> 00:59:26.935 sequencing, but also with methylation
NOTE Confidence: 0.9588068

00:59:26.995 --> 00:59:28.835 analysis or other forms of
NOTE Confidence: 0.9588068

00:59:28.835 --> 00:59:30.755 epigenetic analysis. But I think
NOTE Confidence: 0.9588068

00:59:30.755 --> 00:59:32.195 that you are absolutely correct.
NOTE Confidence: 0.9588068

00:59:32.195 --> 00:59:33.315 This is a direction that
NOTE Confidence: 0.9588068

00:59:33.315 --> 00:59:34.135 we are following.
NOTE Confidence: 0.99373955

00:59:34.435 --> 00:59:35.155 I don't know if we
NOTE Confidence: 0.99373955

00:59:35.155 --> 00:59:36.755 will get there sooner or
NOTE Confidence: 0.99373955

00:59:36.755 --> 00:59:37.975 later, but we will.
NOTE Confidence: 0.66482544

00:59:39.579 --> 00:59:40.859 The focus of the non
NOTE Confidence: 0.66482544

00:59:40.859 --> 00:59:42.160 coding gene, right?

NOTE Confidence: 0.88879395
00:59:42.540 --> 00:59:43.599 Yeah. Exactly.
NOTE Confidence: 0.78175324
00:59:44.300 --> 00:59:46.060 Yeah. Junk. Yes. They they
NOTE Confidence: 0.78175324
00:59:46.060 --> 00:59:47.820 so called junk. Yeah. They
NOTE Confidence: 0.78175324
00:59:47.820 --> 00:59:49.599 hit the or chromatin. Chromatin.
NOTE Confidence: 0.93774414
00:59:50.060 --> 00:59:50.560 Yes.
NOTE Confidence: 0.9471959
01:00:03.035 --> 01:00:04.410 Is there any progress in
NOTE Confidence: 0.9471959
01:00:04.410 --> 01:00:06.010 that? Well, I think there
NOTE Confidence: 0.9471959
01:00:06.010 --> 01:00:07.390 is progress in terms
NOTE Confidence: 0.9970703
01:00:09.530 --> 01:00:10.270 of stopping
NOTE Confidence: 0.96202505
01:00:11.050 --> 01:00:12.890 the growth development in certain
NOTE Confidence: 0.96202505
01:00:12.890 --> 01:00:14.890 examples, but not in many
NOTE Confidence: 0.96202505
01:00:14.890 --> 01:00:15.072 examples.
NOTE Confidence: 0.8046875
01:00:17.734 --> 01:00:18.954 You you cannot change the
NOTE Confidence: 0.8735657
01:00:19.255 --> 01:00:20.954 genetic composition with targeted therapy.
NOTE Confidence: 0.8735657
01:00:21.095 --> 01:00:21.994 You can invalidate
NOTE Confidence: 0.8409424

01:00:22.535 --> 01:00:24.474 or annulate the effect of
NOTE Confidence: 0.890625

01:00:24.775 --> 01:00:25.974 a given gene in in
NOTE Confidence: 0.890625

01:00:25.974 --> 01:00:26.474 general.
NOTE Confidence: 0.89367676

01:00:27.095 --> 01:00:29.035 There is genetic therapy
NOTE Confidence: 0.9734941

01:00:29.630 --> 01:00:31.650 that is changing the genome
NOTE Confidence: 0.9734941

01:00:31.790 --> 01:00:33.730 in certain conditions, not necessarily
NOTE Confidence: 0.9734941

01:00:33.870 --> 01:00:34.370 neoplastic,
NOTE Confidence: 0.94097155

01:00:35.230 --> 01:00:37.310 Duchenne dystrophy, for example. There
NOTE Confidence: 0.94097155

01:00:37.310 --> 01:00:39.070 are examples of genetic approaches
NOTE Confidence: 0.94097155

01:00:39.070 --> 01:00:41.650 that correct the genetic abnormality
NOTE Confidence: 0.94097155

01:00:41.870 --> 01:00:43.490 and therefore will prevent
NOTE Confidence: 0.7998047

01:00:44.175 --> 01:00:44.575 the,
NOTE Confidence: 0.98438084

01:00:44.975 --> 01:00:46.735 continuation of the disorder. But
NOTE Confidence: 0.98438084

01:00:46.735 --> 01:00:48.175 Farzana may be able to
NOTE Confidence: 0.98438084

01:00:48.175 --> 01:00:49.475 respond to that question.
NOTE Confidence: 0.7335744

01:00:49.775 --> 01:00:50.975 Yeah. So I think the

NOTE Confidence: 0.7335744
01:00:51.055 --> 01:00:52.435 at the moment, it seems
NOTE Confidence: 0.7335744
01:00:52.495 --> 01:00:54.335 like diagnosis symptoms, etcetera. There
NOTE Confidence: 0.7335744
01:00:54.335 --> 01:00:55.075 is no
NOTE Confidence: 0.77663165
01:00:55.855 --> 01:00:57.135 target with therapy. But I
NOTE Confidence: 0.77663165
01:00:57.135 --> 01:00:58.575 think the reason to diagnose
NOTE Confidence: 0.77663165
01:00:58.575 --> 01:00:59.395 them early
NOTE Confidence: 0.6165039
01:00:59.840 --> 01:01:01.460 is that we are screening
NOTE Confidence: 0.6165039
01:01:01.600 --> 01:01:03.300 algorithms and we should actually
NOTE Confidence: 0.8164095
01:01:03.760 --> 01:01:05.360 the type three, for example,
NOTE Confidence: 0.8164095
01:01:05.360 --> 01:01:07.360 PPD is the individual treat.
NOTE Confidence: 0.8164095
01:01:07.360 --> 01:01:08.000 But if you have a
NOTE Confidence: 0.8164095
01:01:08.000 --> 01:01:09.440 type one hour, you can
NOTE Confidence: 0.8164095
01:01:09.440 --> 01:01:10.800 actually resect it and go
NOTE Confidence: 0.8164095
01:01:10.800 --> 01:01:12.160 down. So I think all
NOTE Confidence: 0.8164095
01:01:12.160 --> 01:01:13.600 these screening matches that. In
NOTE Confidence: 0.8164095

01:01:13.600 --> 01:01:14.295 fact, they
NOTE Confidence: 0.92285156
01:02:46.240 --> 01:02:46.820 I think
NOTE Confidence: 0.95761716
01:02:47.520 --> 01:02:48.740 I think it will happen,
NOTE Confidence: 0.9968262
01:02:49.280 --> 01:02:50.900 but it's not happening yet.
NOTE Confidence: 0.9968262
01:02:51.120 --> 01:02:51.620 Okay?
NOTE Confidence: 0.950331
01:03:03.015 --> 01:03:05.015 Well, thank you again to
NOTE Confidence: 0.950331
01:03:05.015 --> 01:03:06.315 all of you, and
NOTE Confidence: 0.99072266
01:03:07.015 --> 01:03:07.835 much appreciated.
NOTE Confidence: 0.7829726
01:21:04.265 --> 01:21:05.785 Yes. The the form is
NOTE Confidence: 0.7829726
01:21:05.785 --> 01:21:07.385 still in the post, you
NOTE Confidence: 0.7829726
01:21:07.385 --> 01:21:09.064 know, the In person How
NOTE Confidence: 0.7829726
01:21:09.064 --> 01:21:09.885 you going, sir?
NOTE Confidence: 0.85319006
01:21:10.265 --> 01:21:11.385 I like it. I'm going
NOTE Confidence: 0.85319006
01:21:11.385 --> 01:21:12.425 to dinner with you. Yeah.
NOTE Confidence: 0.85319006
01:21:12.425 --> 01:21:13.545 I know. I'm looking forward
NOTE Confidence: 0.85319006
01:21:13.545 --> 01:21:14.810 to having dinner with you.

NOTE Confidence: 0.85319006
01:21:14.810 --> 01:21:15.850 That's I have a friend.
NOTE Confidence: 0.85319006
01:21:15.850 --> 01:21:17.050 We're going to O'leo's, I
NOTE Confidence: 0.85319006
01:21:17.050 --> 01:21:18.729 guess. Yeah. Yeah. I have
NOTE Confidence: 0.85319006
01:21:18.729 --> 01:21:19.770 to say hi to the
NOTE Confidence: 0.85319006
01:21:19.770 --> 01:21:21.370 other half. Hi. How are
NOTE Confidence: 0.85319006
01:21:21.370 --> 01:21:21.870 you?
NOTE Confidence: 0.46899414
01:21:22.490 --> 01:21:22.990 Hi.
NOTE Confidence: 0.71979725
01:21:23.610 --> 01:21:24.330 I don't know if I
NOTE Confidence: 0.71979725
01:21:24.330 --> 01:21:25.689 like Hi. Super good. Nice
NOTE Confidence: 0.71979725
01:21:25.689 --> 01:21:26.261 to meet you. Yeah. Yeah.
NOTE Confidence: 0.71979725
01:21:26.261 --> 01:21:27.070 Yeah. Yeah. Yeah. Yeah.
NOTE Confidence: 0.73950195
01:21:36.695 --> 01:21:37.195 Okay.
NOTE Confidence: 0.86151993
01:21:41.650 --> 01:21:43.170 I I forgot. That's that's
NOTE Confidence: 0.86151993
01:21:43.170 --> 01:21:44.210 why. Yeah. No. No. I
NOTE Confidence: 0.86151993
01:21:44.210 --> 01:21:45.330 talk about this this,
NOTE Confidence: 0.5984375

01:21:46.530 --> 01:21:47.590 as well as I'm still
NOTE Confidence: 0.7919922

01:21:47.890 --> 01:21:47.970 at
NOTE Confidence: 0.6496718

01:21:49.490 --> 01:21:51.053 I know I know. I
NOTE Confidence: 0.6496718

01:21:51.056 --> 01:21:52.390 know. I know. Yeah.
NOTE Confidence: 0.8972168

01:22:00.285 --> 01:22:00.785 Okay.
NOTE Confidence: 0.821696

01:22:01.485 --> 01:22:02.765 Okay. I think,
NOTE Confidence: 0.95288086

01:22:03.165 --> 01:22:03.905 good afternoon,
NOTE Confidence: 0.99658203

01:22:04.925 --> 01:22:05.425 everyone.
NOTE Confidence: 0.9902344

01:22:06.045 --> 01:22:07.185 Welcome to
NOTE Confidence: 0.7944336

01:22:07.965 --> 01:22:08.625 your pathology
NOTE Confidence: 0.9194336

01:22:10.979 --> 01:22:12.520 ground runs. Have you ever
NOTE Confidence: 0.9194336

01:22:12.739 --> 01:22:13.239 wondered
NOTE Confidence: 0.94873047

01:22:13.939 --> 01:22:15.800 why children get a malignant
NOTE Confidence: 0.94873047

01:22:15.860 --> 01:22:17.219 tumor at such a young
NOTE Confidence: 0.94873047

01:22:17.219 --> 01:22:17.719 age?
NOTE Confidence: 0.9815674

01:22:18.099 --> 01:22:19.460 Today, you're going to hear

NOTE Confidence: 0.9815674
01:22:19.460 --> 01:22:20.439 from the expert,
NOTE Confidence: 0.9175781
01:22:20.820 --> 01:22:22.760 get some answers from him.
NOTE Confidence: 0.8996094
01:22:23.625 --> 01:22:26.045 So it's my great honors
NOTE Confidence: 0.85217285
01:22:26.425 --> 01:22:28.845 and pleasure to introduce today's
NOTE Confidence: 0.85217285
01:22:28.905 --> 01:22:29.965 squad run speaker,
NOTE Confidence: 0.36987305
01:22:30.665 --> 01:22:31.165 doctor
NOTE Confidence: 0.97924805
01:22:31.785 --> 01:22:32.925 Miguel Reyes.
NOTE Confidence: 0.9156189
01:22:34.185 --> 01:22:36.104 So probably doctor Reyes don't
NOTE Confidence: 0.9156189
01:22:36.104 --> 01:22:37.085 need a introduction.
NOTE Confidence: 0.86360675
01:22:38.200 --> 01:22:38.700 He's,
NOTE Confidence: 0.95419544
01:22:39.240 --> 01:22:41.320 you know, has friends here
NOTE Confidence: 0.95419544
01:22:41.320 --> 01:22:43.100 with Yale for many years,
NOTE Confidence: 0.95419544
01:22:43.400 --> 01:22:44.220 has a great
NOTE Confidence: 0.7607422
01:22:44.600 --> 01:22:45.100 friend.
NOTE Confidence: 0.99104816
01:22:45.800 --> 01:22:47.020 So just briefly,
NOTE Confidence: 0.9970703

01:22:47.880 --> 01:22:48.700 doctor Reyes
NOTE Confidence: 0.96191406

01:22:49.080 --> 01:22:49.820 is currently
NOTE Confidence: 0.9973958

01:22:50.120 --> 01:22:51.420 a clinical professor
NOTE Confidence: 0.87280273

01:22:52.425 --> 01:22:53.165 and pediatric
NOTE Confidence: 0.87556964

01:22:53.865 --> 01:22:56.125 and the developmental pathology division
NOTE Confidence: 0.87556964

01:22:56.185 --> 01:22:56.685 chief
NOTE Confidence: 0.8731079

01:22:57.305 --> 01:22:58.685 in the department of pathology
NOTE Confidence: 0.8731079

01:22:58.745 --> 01:23:00.205 and laboratory medicine
NOTE Confidence: 1

01:23:00.905 --> 01:23:01.405 at
NOTE Confidence: 0.9555664

01:23:02.985 --> 01:23:04.205 University of Miami,
NOTE Confidence: 0.95458984

01:23:05.705 --> 01:23:07.005 Miller School of Medicine.
NOTE Confidence: 0.99121094

01:23:08.450 --> 01:23:08.950 Doctor
NOTE Confidence: 0.38671875

01:23:09.490 --> 01:23:09.990 Reyes
NOTE Confidence: 0.98339844

01:23:10.450 --> 01:23:12.310 completed his medical education
NOTE Confidence: 0.9729004

01:23:12.930 --> 01:23:13.830 in National
NOTE Confidence: 0.99902344

01:23:14.370 --> 01:23:14.870 Autonomous

NOTE Confidence: 1
01:23:15.890 --> 01:23:16.390 University
NOTE Confidence: 0.99902344
01:23:16.930 --> 01:23:17.750 of Mexico
NOTE Confidence: 0.8300238
01:23:18.450 --> 01:23:19.910 in nineteen eighties,
NOTE Confidence: 0.93330383
01:23:21.155 --> 01:23:23.635 followed by residency training and
NOTE Confidence: 0.93330383
01:23:23.795 --> 01:23:25.415 in anatomic massage
NOTE Confidence: 0.99609375
01:23:25.875 --> 01:23:26.375 there.
NOTE Confidence: 0.99869794
01:23:27.395 --> 01:23:28.615 After several years
NOTE Confidence: 0.9814453
01:23:29.075 --> 01:23:31.475 working as attending physician in
NOTE Confidence: 0.9814453
01:23:31.475 --> 01:23:32.535 National Institute
NOTE Confidence: 0.9953613
01:23:33.315 --> 01:23:34.055 of Pediatrics,
NOTE Confidence: 0.84521484
01:23:34.960 --> 01:23:36.719 he decided come to United
NOTE Confidence: 0.84521484
01:23:36.719 --> 01:23:37.219 States
NOTE Confidence: 0.75683594
01:23:38.079 --> 01:23:38.739 to continue
NOTE Confidence: 0.9491374
01:23:39.040 --> 01:23:40.500 his medical careers.
NOTE Confidence: 0.9369507
01:23:41.920 --> 01:23:43.860 Doctor. Reyes got his pediatric
NOTE Confidence: 0.9369507

01:23:43.920 --> 01:23:45.540 pathology fellowship training
NOTE Confidence: 0.97094727

01:23:45.920 --> 01:23:46.659 in nineteen
NOTE Confidence: 0.9975586

01:23:47.040 --> 01:23:47.780 ninety four
NOTE Confidence: 0.9876709

01:23:48.555 --> 01:23:51.035 from Children's Memorial Hospital of
NOTE Confidence: 0.9876709

01:23:51.035 --> 01:23:51.535 Northwestern
NOTE Confidence: 0.7458496

01:23:51.995 --> 01:23:52.495 University.
NOTE Confidence: 0.962972

01:23:54.074 --> 01:23:55.215 After the fellowship,
NOTE Confidence: 0.99902344

01:23:56.074 --> 01:23:57.455 he joined Yale
NOTE Confidence: 0.9983259

01:23:57.995 --> 01:23:59.995 as a faculty member in
NOTE Confidence: 0.9983259

01:23:59.995 --> 01:24:01.055 our department
NOTE Confidence: 0.75683594

01:24:02.155 --> 01:24:02.655 and
NOTE Confidence: 0.8338216

01:24:04.020 --> 01:24:05.400 raise his rank
NOTE Confidence: 0.92647296

01:24:05.939 --> 01:24:08.020 from assistant professor to full
NOTE Confidence: 0.92647296

01:24:08.020 --> 01:24:08.520 professors.
NOTE Confidence: 0.97265625

01:24:10.740 --> 01:24:13.400 So after spending fourteen years
NOTE Confidence: 1

01:24:13.700 --> 01:24:14.200 in

NOTE Confidence: 0.99658203
01:24:14.660 --> 01:24:15.160 Yale,
NOTE Confidence: 0.99902344
01:24:15.860 --> 01:24:16.360 he
NOTE Confidence: 0.9977214
01:24:16.660 --> 01:24:18.360 relocated to Pittsburgh
NOTE Confidence: 0.94033206
01:24:19.695 --> 01:24:21.715 to become the chief pathologist
NOTE Confidence: 0.99125975
01:24:22.094 --> 01:24:24.354 in Children's Hospital of Pittsburgh
NOTE Confidence: 0.8031006
01:24:25.935 --> 01:24:27.155 University, Pittsburgh
NOTE Confidence: 0.95874023
01:24:28.574 --> 01:24:30.675 until twenty twenty four.
NOTE Confidence: 0.87561035
01:24:32.180 --> 01:24:33.960 Doctor. Reyes is a world
NOTE Confidence: 0.87561035
01:24:34.180 --> 01:24:34.680 renowned
NOTE Confidence: 0.9488119
01:24:35.460 --> 01:24:36.520 pediatric pathologist,
NOTE Confidence: 0.96875
01:24:37.380 --> 01:24:37.880 has
NOTE Confidence: 0.7732747
01:24:38.180 --> 01:24:40.040 made significant contribution
NOTE Confidence: 0.9199707
01:24:40.820 --> 01:24:43.240 to the pediatric pathologist communities,
NOTE Confidence: 0.9973958
01:24:43.939 --> 01:24:45.640 including clinical practice,
NOTE Confidence: 1
01:24:46.715 --> 01:24:47.215 translational
NOTE Confidence: 0.8544922

01:24:47.515 --> 01:24:48.015 research,
NOTE Confidence: 0.984592
01:24:49.275 --> 01:24:50.814 and medical educations.
NOTE Confidence: 0.9607476
01:24:53.195 --> 01:24:55.615 He was the past president
NOTE Confidence: 0.9607476
01:24:55.915 --> 01:24:58.335 of International Pediatric Massage
NOTE Confidence: 0.9951172
01:24:58.715 --> 01:24:59.215 Associations
NOTE Confidence: 0.9121094
01:25:00.540 --> 01:25:01.280 and the Society
NOTE Confidence: 0.98879665
01:25:02.540 --> 01:25:04.000 of Pediatric Massage.
NOTE Confidence: 0.58691406
01:25:05.900 --> 01:25:06.960 Doctor. Reyes
NOTE Confidence: 0.8442383
01:25:07.420 --> 01:25:07.920 published
NOTE Confidence: 0.9459054
01:25:08.619 --> 01:25:10.540 more than three hundred peer
NOTE Confidence: 0.9459054
01:25:10.540 --> 01:25:11.599 review articles,
NOTE Confidence: 0.69750977
01:25:12.619 --> 01:25:13.119 books,
NOTE Confidence: 0.87353516
01:25:13.579 --> 01:25:15.280 books, and book chapters.
NOTE Confidence: 0.96028644
01:25:18.255 --> 01:25:20.115 So he is the standing
NOTE Confidence: 0.96028644
01:25:20.415 --> 01:25:20.915 editors
NOTE Confidence: 0.93481445
01:25:21.295 --> 01:25:22.675 of the WHO classification

NOTE Confidence: 0.970459
01:25:23.535 --> 01:25:24.115 of tumors
NOTE Confidence: 0.9267578
01:25:24.895 --> 01:25:25.395 editorial
NOTE Confidence: 0.6911621
01:25:25.695 --> 01:25:26.195 board.
NOTE Confidence: 0.63346356
01:25:26.655 --> 01:25:27.475 And he was
NOTE Confidence: 0.7409668
01:25:28.095 --> 01:25:28.915 the expert
NOTE Confidence: 0.9970703
01:25:29.295 --> 01:25:29.795 editor
NOTE Confidence: 0.9815267
01:25:30.660 --> 01:25:32.820 for the newly released WHO
NOTE Confidence: 0.9815267
01:25:32.820 --> 01:25:33.320 classification
NOTE Confidence: 0.9921875
01:25:33.780 --> 01:25:34.520 of tumor
NOTE Confidence: 0.7246094
01:25:35.300 --> 01:25:35.800 pediatric
NOTE Confidence: 0.99853516
01:25:36.739 --> 01:25:37.239 tumors.
NOTE Confidence: 0.8965149
01:25:38.420 --> 01:25:40.120 So although, you know,
NOTE Confidence: 0.9802246
01:25:40.500 --> 01:25:42.120 the current WHO classification
NOTE Confidence: 0.99609375
01:25:42.500 --> 01:25:43.000 tumor
NOTE Confidence: 0.96435547
01:25:43.540 --> 01:25:44.660 already in his
NOTE Confidence: 0.86499023

01:25:45.275 --> 01:25:47.135 its fifth editions.
NOTE Confidence: 0.9563802

01:25:48.235 --> 01:25:49.614 The pediatric tumors
NOTE Confidence: 0.98132324

01:25:50.314 --> 01:25:51.614 is the first time
NOTE Confidence: 0.9682617

01:25:51.915 --> 01:25:53.534 WHO has including
NOTE Confidence: 0.9583333

01:25:53.915 --> 01:25:55.215 a separate volume
NOTE Confidence: 0.9892578

01:25:56.074 --> 01:25:57.695 on the pediatric tumors.
NOTE Confidence: 0.9243164

01:25:59.449 --> 01:26:01.469 There's no doubt this publication
NOTE Confidence: 0.9889323

01:26:01.929 --> 01:26:03.150 will help improving
NOTE Confidence: 0.9968262

01:26:04.090 --> 01:26:04.909 the standardization
NOTE Confidence: 0.99902344

01:26:05.689 --> 01:26:06.429 of pediatric
NOTE Confidence: 0.99853516

01:26:06.810 --> 01:26:08.030 tumor diagnosis
NOTE Confidence: 0.982666

01:26:08.810 --> 01:26:09.469 and facilitate
NOTE Confidence: 0.74072266

01:26:10.489 --> 01:26:11.310 the translate
NOTE Confidence: 0.9977214

01:26:12.375 --> 01:26:13.914 translation of diagnostic
NOTE Confidence: 0.9975586

01:26:14.215 --> 01:26:15.434 research into
NOTE Confidence: 1

01:26:15.974 --> 01:26:16.474 practice

NOTE Confidence: 0.99365234
01:26:17.014 --> 01:26:17.514 worldwide.
NOTE Confidence: 0.975708
01:26:18.695 --> 01:26:20.635 So without further ado,
NOTE Confidence: 0.8231934
01:26:21.255 --> 01:26:23.594 please join me, Wolfgang. Reyes.
NOTE Confidence: 0.99853516
01:26:31.139 --> 01:26:32.599 Well, thank you very much
NOTE Confidence: 0.8342285
01:26:33.460 --> 01:26:34.199 for being
NOTE Confidence: 0.8026123
01:26:34.900 --> 01:26:36.360 first for inviting me,
NOTE Confidence: 0.9053955
01:26:36.900 --> 01:26:37.540 then for,
NOTE Confidence: 0.9142456
01:26:38.020 --> 01:26:40.420 presenting me as in such
NOTE Confidence: 0.9142456
01:26:40.420 --> 01:26:41.560 a kindly manner.
NOTE Confidence: 0.96972656
01:26:42.975 --> 01:26:45.055 I recognize and have a
NOTE Confidence: 0.96972656
01:26:45.055 --> 01:26:45.535 lot of,
NOTE Confidence: 0.984375
01:26:46.895 --> 01:26:47.395 fun
NOTE Confidence: 0.99790734
01:26:48.175 --> 01:26:49.375 to see many of my
NOTE Confidence: 0.99790734
01:26:49.375 --> 01:26:50.195 former trainees,
NOTE Confidence: 0.9975586
01:26:50.975 --> 01:26:51.475 colleagues,
NOTE Confidence: 0.9765625

01:26:51.854 --> 01:26:52.354 friends,
NOTE Confidence: 0.9996745

01:26:53.695 --> 01:26:54.995 and people that
NOTE Confidence: 0.9717

01:26:55.340 --> 01:26:57.179 taught me how to become
NOTE Confidence: 0.9717

01:26:57.179 --> 01:26:58.479 an academic pathologist.
NOTE Confidence: 0.73051757

01:26:59.099 --> 01:27:01.679 There's Costa, doctor Morrow here,
NOTE Confidence: 0.99316406

01:27:02.300 --> 01:27:02.800 and
NOTE Confidence: 0.75878906

01:27:03.340 --> 01:27:04.400 David here.
NOTE Confidence: 0.9909668

01:27:05.019 --> 01:27:05.840 We interacted
NOTE Confidence: 0.90563965

01:27:06.300 --> 01:27:07.500 the other day with here.
NOTE Confidence: 0.90563965

01:27:07.500 --> 01:27:09.260 We interacted with them many
NOTE Confidence: 0.90563965

01:27:09.260 --> 01:27:09.900 years, and,
NOTE Confidence: 0.977948

01:27:10.854 --> 01:27:11.895 it has been an honor
NOTE Confidence: 0.977948

01:27:11.895 --> 01:27:12.935 for me. This is really
NOTE Confidence: 0.977948

01:27:12.935 --> 01:27:14.534 my academic home, so I
NOTE Confidence: 0.977948

01:27:14.534 --> 01:27:16.395 am very glad to return.
NOTE Confidence: 0.98344725

01:27:18.295 --> 01:27:20.695 Today, I will present something

NOTE Confidence: 0.98344725
01:27:20.695 --> 01:27:22.295 that is different from, the
NOTE Confidence: 0.98344725
01:27:22.295 --> 01:27:23.994 previous conference that I presented
NOTE Confidence: 0.98344725
01:27:24.135 --> 01:27:26.074 here about eight years ago.
NOTE Confidence: 0.9589844
01:27:26.420 --> 01:27:28.340 Today, we will, talk about
NOTE Confidence: 0.9589844
01:27:28.340 --> 01:27:28.840 how
NOTE Confidence: 0.9641462
01:27:29.300 --> 01:27:30.820 looking at the slice in
NOTE Confidence: 0.9641462
01:27:30.820 --> 01:27:32.040 pediatric lesions
NOTE Confidence: 0.9984375
01:27:32.420 --> 01:27:33.800 can allow you to identify
NOTE Confidence: 0.9958496
01:27:35.140 --> 01:27:36.040 cancer predisposition
NOTE Confidence: 0.98501587
01:27:36.420 --> 01:27:38.100 syndromes. This is a topic
NOTE Confidence: 0.98501587
01:27:38.100 --> 01:27:38.660 that I have,
NOTE Confidence: 0.86865234
01:27:40.405 --> 01:27:40.905 cultivated,
NOTE Confidence: 1
01:27:41.764 --> 01:27:42.264 extensively
NOTE Confidence: 0.9973389
01:27:42.564 --> 01:27:43.844 in the last few years.
NOTE Confidence: 0.9973389
01:27:43.844 --> 01:27:45.945 How do I advance this?
NOTE Confidence: 0.93263245

01:27:52.210 --> 01:27:53.570 Oh, thank you very much.
NOTE Confidence: 0.93263245

01:27:53.570 --> 01:27:54.710 Didn't see that.
NOTE Confidence: 0.9925537

01:27:55.650 --> 01:27:57.270 So let's move on.
NOTE Confidence: 0.9389893

01:27:59.570 --> 01:28:01.030 I have nothing to declare,
NOTE Confidence: 0.96904296

01:28:02.210 --> 01:28:03.989 and, this is the outline,
NOTE Confidence: 0.99532646

01:28:05.335 --> 01:28:06.854 and we will go directly
NOTE Confidence: 0.99532646

01:28:06.854 --> 01:28:07.594 into matters.
NOTE Confidence: 0.9719238

01:28:08.614 --> 01:28:09.895 At the beginning of last
NOTE Confidence: 0.9719238

01:28:09.895 --> 01:28:12.635 year, the official's archive journal,
NOTE Confidence: 0.9719238

01:28:12.695 --> 01:28:14.235 the oldest journal of pathology
NOTE Confidence: 0.9719238

01:28:14.454 --> 01:28:15.195 in the world,
NOTE Confidence: 0.92729384

01:28:16.054 --> 01:28:18.054 published a special issue in
NOTE Confidence: 0.92729384

01:28:18.054 --> 01:28:20.000 which, my colleague from Madrid,
NOTE Confidence: 0.92729384

01:28:20.000 --> 01:28:21.920 Doctor. Isabel Colmenero, the chief
NOTE Confidence: 0.92729384

01:28:21.920 --> 01:28:24.000 pathologist at Hospital del Nino
NOTE Confidence: 0.92729384

01:28:24.000 --> 01:28:25.540 Jesus in Madrid,

NOTE Confidence: 0.9988839
01:28:26.400 --> 01:28:27.840 and I put together a
NOTE Confidence: 0.9988839
01:28:27.840 --> 01:28:28.820 collection of
NOTE Confidence: 0.9478469
01:28:29.200 --> 01:28:30.720 twelve papers, ten of twelve
NOTE Confidence: 0.9478469
01:28:30.720 --> 01:28:32.915 papers, and an editorial. We
NOTE Confidence: 0.9478469
01:28:32.915 --> 01:28:34.275 invited all the authors that
NOTE Confidence: 0.9478469
01:28:34.275 --> 01:28:36.435 contributed to this special issue
NOTE Confidence: 0.9478469
01:28:36.435 --> 01:28:36.935 highlighting
NOTE Confidence: 0.94311523
01:28:37.555 --> 01:28:37.955 the,
NOTE Confidence: 0.9792259
01:28:38.435 --> 01:28:40.375 challenges and novelties in pediatric
NOTE Confidence: 0.9792259
01:28:40.435 --> 01:28:42.115 pathology. I invite you to
NOTE Confidence: 0.9792259
01:28:42.115 --> 01:28:43.715 review the special issue because
NOTE Confidence: 0.9792259
01:28:43.715 --> 01:28:45.715 it contains interesting information that
NOTE Confidence: 0.9792259
01:28:45.715 --> 01:28:46.775 is mostly
NOTE Confidence: 0.99384767
01:28:47.389 --> 01:28:49.650 missed by the adult pathology,
NOTE Confidence: 0.93570966
01:28:50.670 --> 01:28:52.530 sphere, the adult adult pathology
NOTE Confidence: 0.93570966

01:28:52.750 --> 01:28:53.250 circles.
NOTE Confidence: 0.9926758
01:28:54.349 --> 01:28:55.090 In this
NOTE Confidence: 0.73950195
01:28:56.429 --> 01:28:56.929 issue,
NOTE Confidence: 1
01:28:57.229 --> 01:28:58.369 one of the papers
NOTE Confidence: 0.99786377
01:28:58.670 --> 01:29:00.685 was specifically dedicated to the
NOTE Confidence: 0.99786377
01:29:00.685 --> 01:29:01.985 pediatric cancer predisposition
NOTE Confidence: 0.9983724
01:29:02.285 --> 01:29:02.785 syndromes
NOTE Confidence: 0.94295245
01:29:03.165 --> 01:29:04.385 in the, outside,
NOTE Confidence: 0.97324216
01:29:04.844 --> 01:29:06.785 of the central nervous system.
NOTE Confidence: 0.9894456
01:29:07.245 --> 01:29:08.765 And, I also invite you
NOTE Confidence: 0.9894456
01:29:08.765 --> 01:29:10.045 to read this paper because
NOTE Confidence: 0.9894456
01:29:10.045 --> 01:29:11.025 this will summarize
NOTE Confidence: 0.99934894
01:29:11.405 --> 01:29:12.445 what I am going to
NOTE Confidence: 0.99934894
01:29:12.445 --> 01:29:13.745 present to you today.
NOTE Confidence: 0.9498291
01:29:16.679 --> 01:29:18.139 K. It's not advancing.
NOTE Confidence: 0.96931154
01:29:18.439 --> 01:29:20.280 Okay. Here. So how do

NOTE Confidence: 0.96931154

01:29:20.280 --> 01:29:22.619 we define cancer predisposition syndromes?

NOTE Confidence: 0.9702497

01:29:23.479 --> 01:29:25.800 Heritable condition that increases the

NOTE Confidence: 0.9702497

01:29:25.800 --> 01:29:27.559 child's risk of developing cancer

NOTE Confidence: 0.9702497

01:29:27.559 --> 01:29:29.020 due to general mutations

NOTE Confidence: 0.89282227

01:29:29.585 --> 01:29:30.485 gene regulating

NOTE Confidence: 0.92396766

01:29:30.864 --> 01:29:32.784 cell growth, DNA repair, or

NOTE Confidence: 0.92396766

01:29:32.784 --> 01:29:34.705 genomic integrity. That's in general

NOTE Confidence: 0.92396766

01:29:34.705 --> 01:29:35.844 where a cancer predisposition

NOTE Confidence: 0.9408514

01:29:36.145 --> 01:29:37.344 syndrome is. And, of course,

NOTE Confidence: 0.9408514

01:29:37.344 --> 01:29:38.965 this affects not only children,

NOTE Confidence: 0.9408514

01:29:39.185 --> 01:29:39.685 but

NOTE Confidence: 0.8884033

01:29:40.145 --> 01:29:42.180 adults too. But adults affected

NOTE Confidence: 0.8884033

01:29:42.240 --> 01:29:44.180 with these conditions bring these

NOTE Confidence: 0.94226074

01:29:44.560 --> 01:29:46.020 conditions since they are

NOTE Confidence: 0.88740236

01:29:46.560 --> 01:29:48.900 children. So we pediatric pathologists

NOTE Confidence: 0.9756557

01:29:49.280 --> 01:29:51.120 are at the specific point
NOTE Confidence: 0.9756557

01:29:51.120 --> 01:29:51.860 to identify
NOTE Confidence: 0.9758301

01:29:52.160 --> 01:29:53.300 these entities
NOTE Confidence: 0.97920734

01:29:53.600 --> 01:29:55.814 early, early enough to intervene
NOTE Confidence: 0.97920734

01:29:56.114 --> 01:29:58.114 and modify the outcomes of
NOTE Confidence: 0.97920734

01:29:58.114 --> 01:29:59.014 these patients.
NOTE Confidence: 0.92838544

01:29:59.635 --> 01:30:01.175 They are, representing,
NOTE Confidence: 0.5283203

01:30:02.434 --> 01:30:02.934 in
NOTE Confidence: 0.81693524

01:30:03.474 --> 01:30:06.375 inherited risk, usually autosomal dominant,
NOTE Confidence: 0.81693524

01:30:06.675 --> 01:30:07.175 occasionally
NOTE Confidence: 0.7265625

01:30:07.630 --> 01:30:08.290 de novo,
NOTE Confidence: 0.98779297

01:30:08.590 --> 01:30:10.590 and the tumor often appears
NOTE Confidence: 0.98779297

01:30:10.590 --> 01:30:12.430 at younger ages than, general
NOTE Confidence: 0.98779297

01:30:12.430 --> 01:30:13.870 population. The same type of
NOTE Confidence: 0.98779297

01:30:13.870 --> 01:30:14.910 tumor that you see in
NOTE Confidence: 0.98779297

01:30:14.910 --> 01:30:16.270 adults when it appears in

NOTE Confidence: 0.98779297

01:30:16.270 --> 01:30:17.550 a child, it may be

NOTE Confidence: 0.98779297

01:30:17.550 --> 01:30:19.070 the first clue for you

NOTE Confidence: 0.98779297

01:30:19.070 --> 01:30:20.350 to look for a cancer

NOTE Confidence: 0.98779297

01:30:20.350 --> 01:30:20.850 predisposition

NOTE Confidence: 0.9995117

01:30:21.229 --> 01:30:21.729 syndrome.

NOTE Confidence: 0.9694824

01:30:22.594 --> 01:30:24.275 Multiple of these, many of

NOTE Confidence: 0.9694824

01:30:24.275 --> 01:30:25.814 these tumors are rare

NOTE Confidence: 0.992806

01:30:26.275 --> 01:30:28.534 and, children with multiple primaries

NOTE Confidence: 0.992806

01:30:28.594 --> 01:30:30.935 or unusual types are specifically

NOTE Confidence: 0.992806

01:30:31.074 --> 01:30:32.534 a red flag to identify.

NOTE Confidence: 0.9776964

01:30:33.155 --> 01:30:34.835 The family history may show

NOTE Confidence: 0.9776964

01:30:34.835 --> 01:30:37.010 clustering of specific or related

NOTE Confidence: 0.9776964

01:30:37.010 --> 01:30:39.110 causes, but sometimes it doesn't.

NOTE Confidence: 0.9788485

01:30:39.890 --> 01:30:41.970 Approximately ten percent of pediatric

NOTE Confidence: 0.9788485

01:30:41.970 --> 01:30:43.590 cancers are due to germline

NOTE Confidence: 0.9788485

01:30:43.729 --> 01:30:44.229 alterations
NOTE Confidence: 0.83306885

01:30:45.090 --> 01:30:47.110 and early frequent recognition
NOTE Confidence: 0.9967448

01:30:47.489 --> 01:30:48.390 is a key
NOTE Confidence: 0.73657227

01:30:49.115 --> 01:30:49.615 to,
NOTE Confidence: 0.80997723

01:30:51.035 --> 01:30:52.335 provide adequate surveillance,
NOTE Confidence: 0.9821167

01:30:52.635 --> 01:30:54.575 therapy, and family assessment.
NOTE Confidence: 0.96189374

01:30:55.275 --> 01:30:57.195 So the point that I
NOTE Confidence: 0.96189374

01:30:57.195 --> 01:30:58.795 will emphasize today is that
NOTE Confidence: 0.96189374

01:30:58.795 --> 01:31:00.015 frequently histology
NOTE Confidence: 0.99214476

01:31:00.475 --> 01:31:02.430 provides the first clue, but
NOTE Confidence: 0.99214476

01:31:02.430 --> 01:31:03.390 you just have to keep
NOTE Confidence: 0.99214476

01:31:03.390 --> 01:31:04.830 this in mind. If you
NOTE Confidence: 0.99214476

01:31:04.830 --> 01:31:06.610 don't, you'll miss it.
NOTE Confidence: 0.9941406

01:31:08.190 --> 01:31:08.690 Okay.
NOTE Confidence: 0.9627177

01:31:12.110 --> 01:31:13.965 The pediatric pathologist is then
NOTE Confidence: 0.9627177

01:31:13.965 --> 01:31:15.804 at the crossroads between what

NOTE Confidence: 0.9627177
01:31:15.804 --> 01:31:17.244 is the clinical observation of
NOTE Confidence: 0.9627177
01:31:17.244 --> 01:31:19.005 a patient, the genetics of
NOTE Confidence: 0.9627177
01:31:19.005 --> 01:31:20.385 this patient and evaluating,
NOTE Confidence: 0.9384068
01:31:21.085 --> 01:31:22.764 the lesions that are under
NOTE Confidence: 0.9384068
01:31:22.764 --> 01:31:23.425 the microscope
NOTE Confidence: 0.97942245
01:31:23.965 --> 01:31:25.344 with the risk of increasing
NOTE Confidence: 0.97942245
01:31:25.485 --> 01:31:26.705 developing cancer.
NOTE Confidence: 0.9869908
01:31:27.564 --> 01:31:30.030 In addition to accurately diagnose
NOTE Confidence: 0.9869908
01:31:30.030 --> 01:31:30.770 these entities,
NOTE Confidence: 0.99960935
01:31:31.150 --> 01:31:32.610 we need to be familiarized
NOTE Confidence: 0.9861816
01:31:32.990 --> 01:31:34.930 with the implications of additional,
NOTE Confidence: 1
01:31:36.189 --> 01:31:36.689 seemingly
NOTE Confidence: 0.8766276
01:31:37.950 --> 01:31:38.450 uninteresting,
NOTE Confidence: 0.92874146
01:31:38.830 --> 01:31:40.510 unimportant findings that I will
NOTE Confidence: 0.92874146
01:31:40.510 --> 01:31:41.410 show you today.
NOTE Confidence: 0.9569092

01:31:41.875 --> 01:31:43.735 And clinical and or laboratory
NOTE Confidence: 0.9569092

01:31:43.795 --> 01:31:44.295 findings,
NOTE Confidence: 0.9016113

01:31:45.155 --> 01:31:47.314 that are apparently minor may
NOTE Confidence: 0.9016113

01:31:47.314 --> 01:31:48.295 be of various
NOTE Confidence: 0.99438477

01:31:48.675 --> 01:31:49.895 important significance.
NOTE Confidence: 0.9603749

01:31:50.675 --> 01:31:51.715 This is taken from the
NOTE Confidence: 0.9603749

01:31:51.715 --> 01:31:53.155 paper of the officials archive
NOTE Confidence: 0.9603749

01:31:53.155 --> 01:31:54.514 journal that I mentioned to
NOTE Confidence: 0.9603749

01:31:54.514 --> 01:31:55.875 you. I'm not going to
NOTE Confidence: 0.9603749

01:31:55.875 --> 01:31:57.699 go over every one of
NOTE Confidence: 0.9603749

01:31:57.699 --> 01:31:58.900 these entities. This is just
NOTE Confidence: 0.9603749

01:31:58.900 --> 01:32:00.900 to emphasize that I I
NOTE Confidence: 0.9603749

01:32:00.900 --> 01:32:02.659 would like you to, review
NOTE Confidence: 0.9603749

01:32:02.659 --> 01:32:04.099 the paper, read it, and
NOTE Confidence: 0.9603749

01:32:04.099 --> 01:32:04.500 and,
NOTE Confidence: 0.9794922

01:32:04.900 --> 01:32:07.159 send us any, comments, criticisms,

NOTE Confidence: 0.98235154

01:32:07.540 --> 01:32:09.564 etcetera. There are many can,

NOTE Confidence: 0.98235154

01:32:09.725 --> 01:32:12.045 cancer predisposition syndromes listed here.

NOTE Confidence: 0.98235154

01:32:12.045 --> 01:32:13.245 Obviously, this is just to

NOTE Confidence: 0.98235154

01:32:13.245 --> 01:32:14.525 show you that the paper

NOTE Confidence: 0.98235154

01:32:14.525 --> 01:32:15.265 is long

NOTE Confidence: 0.97924805

01:32:15.645 --> 01:32:16.045 and,

NOTE Confidence: 0.9772949

01:32:16.685 --> 01:32:17.245 full of,

NOTE Confidence: 1

01:32:17.885 --> 01:32:18.385 information

NOTE Confidence: 0.976532

01:32:18.765 --> 01:32:19.725 that I am not going

NOTE Confidence: 0.976532

01:32:19.725 --> 01:32:20.945 to be able to detail

NOTE Confidence: 0.976532

01:32:21.005 --> 01:32:22.365 here. But we are going

NOTE Confidence: 0.976532

01:32:22.365 --> 01:32:22.865 to,

NOTE Confidence: 0.9467076

01:32:23.590 --> 01:32:24.790 start with one of the

NOTE Confidence: 0.9467076

01:32:24.790 --> 01:32:25.610 most common,

NOTE Confidence: 0.99975586

01:32:26.150 --> 01:32:27.910 pediatric tumors that we see

NOTE Confidence: 0.99975586

01:32:27.910 --> 01:32:28.410 here.
NOTE Confidence: 0.9131368
01:32:29.510 --> 01:32:31.110 This is Wilms tumor, also
NOTE Confidence: 0.9131368
01:32:31.110 --> 01:32:32.090 known as nephroblastoma.
NOTE Confidence: 0.9278259
01:32:33.189 --> 01:32:35.030 Nobody really knows what doctor
NOTE Confidence: 0.9278259
01:32:35.030 --> 01:32:36.330 Max Williams described.
NOTE Confidence: 0.9489862
01:32:37.345 --> 01:32:39.345 Bruce Beckwith, the late Bruce
NOTE Confidence: 0.9489862
01:32:39.345 --> 01:32:40.545 Beckwith, a good friend of
NOTE Confidence: 0.9489862
01:32:40.545 --> 01:32:42.005 mine, who was the,
NOTE Confidence: 0.9734049
01:32:42.705 --> 01:32:44.385 official reviewer for the children's
NOTE Confidence: 0.9734049
01:32:44.385 --> 01:32:46.245 oncology group for many years,
NOTE Confidence: 0.92317533
01:32:47.104 --> 01:32:48.865 told me several times that
NOTE Confidence: 0.92317533
01:32:48.865 --> 01:32:50.520 nobody really understood what Max
NOTE Confidence: 0.92317533
01:32:50.600 --> 01:32:52.200 Williams, described, but this is
NOTE Confidence: 0.92317533
01:32:52.200 --> 01:32:53.660 the entity that we recognize
NOTE Confidence: 0.92317533
01:32:53.720 --> 01:32:54.700 as Williams' tumor.
NOTE Confidence: 0.9499512
01:32:55.160 --> 01:32:56.600 A tumor that produces a

NOTE Confidence: 0.9499512
01:32:56.600 --> 01:32:57.100 massive
NOTE Confidence: 0.9698444
01:32:57.640 --> 01:33:00.040 growth inside the kidney with
NOTE Confidence: 0.9698444
01:33:00.040 --> 01:33:01.560 a claw sign on the
NOTE Confidence: 0.9698444
01:33:01.560 --> 01:33:03.645 residual kidney that surrounds it
NOTE Confidence: 0.9698444
01:33:03.805 --> 01:33:04.685 as you can see in
NOTE Confidence: 0.9698444
01:33:04.685 --> 01:33:06.845 this, CT scan. And this
NOTE Confidence: 0.9698444
01:33:06.845 --> 01:33:08.305 is a classic microscopic
NOTE Confidence: 0.96876526
01:33:08.685 --> 01:33:10.125 appearance. I don't use the
NOTE Confidence: 0.96876526
01:33:10.125 --> 01:33:12.365 gross the gross term because
NOTE Confidence: 0.96876526
01:33:12.365 --> 01:33:13.885 it grosses me out. It's
NOTE Confidence: 0.96876526
01:33:13.885 --> 01:33:14.385 recommending
NOTE Confidence: 0.94517297
01:33:14.925 --> 01:33:16.445 it's it's recommended to get
NOTE Confidence: 0.94517297
01:33:16.445 --> 01:33:17.645 rid of the grossing and
NOTE Confidence: 0.94517297
01:33:17.645 --> 01:33:19.660 gross. But, anyway, the microscopic
NOTE Confidence: 0.94517297
01:33:19.960 --> 01:33:21.720 appearance of this tumor is
NOTE Confidence: 0.94517297

01:33:21.720 --> 01:33:22.220 this,
NOTE Confidence: 0.991862

01:33:23.240 --> 01:33:23.740 fleshy,
NOTE Confidence: 0.83443195

01:33:24.920 --> 01:33:27.979 relatively homogeneous, sometimes, vocally hemorrhagic,
NOTE Confidence: 0.950217

01:33:28.680 --> 01:33:30.600 sometimes very hemorrhagic. And then
NOTE Confidence: 0.950217

01:33:30.600 --> 01:33:31.720 you can appreciate that there
NOTE Confidence: 0.950217

01:33:31.720 --> 01:33:33.100 is a little rim of
NOTE Confidence: 0.9946289

01:33:33.465 --> 01:33:34.685 kidney that,
NOTE Confidence: 0.9666748

01:33:35.785 --> 01:33:36.445 is still,
NOTE Confidence: 0.99731445

01:33:37.145 --> 01:33:38.045 preserved there.
NOTE Confidence: 0.9201327

01:33:38.345 --> 01:33:39.865 This is Bruce Beckwith here
NOTE Confidence: 0.9201327

01:33:39.865 --> 01:33:41.064 visiting at Yale. This is
NOTE Confidence: 0.9201327

01:33:41.064 --> 01:33:42.285 me, believe it or not.
NOTE Confidence: 0.9201327

01:33:42.425 --> 01:33:43.945 I still didn't stain my
NOTE Confidence: 0.9201327

01:33:43.945 --> 01:33:44.765 beard white.
NOTE Confidence: 0.9700425

01:33:45.305 --> 01:33:46.665 And, this is a few
NOTE Confidence: 0.9700425

01:33:46.665 --> 01:33:47.945 years later, ten years later,
NOTE Confidence: 0.9700425

01:33:47.945 --> 01:33:49.140 when we met together in
NOTE Confidence: 0.9700425

01:33:49.140 --> 01:33:50.360 Tours, France.
NOTE Confidence: 0.7946715

01:33:51.780 --> 01:33:53.719 This Beckwith William and syndrome
NOTE Confidence: 0.7946715

01:33:53.860 --> 01:33:55.380 and other syndromes such as
NOTE Confidence: 0.7946715

01:33:55.380 --> 01:33:56.760 Wagner, Dennis Trash,
NOTE Confidence: 0.8815918

01:33:57.140 --> 01:33:58.199 Simpson Golami,
NOTE Confidence: 0.8838501

01:33:58.979 --> 01:34:00.199 Behamel, Fraser,
NOTE Confidence: 0.85393965

01:34:00.500 --> 01:34:02.420 Bloom syndrome, Dyson one are
NOTE Confidence: 0.85393965

01:34:02.420 --> 01:34:04.795 associated with Wilms tumors and
NOTE Confidence: 0.85393965

01:34:04.795 --> 01:34:06.635 we will review how they
NOTE Confidence: 0.85393965

01:34:06.635 --> 01:34:08.415 look at, very briefly.
NOTE Confidence: 0.9700702

01:34:08.795 --> 01:34:11.295 Triphasic Wilms tumors with epithelial
NOTE Confidence: 0.9700702

01:34:11.594 --> 01:34:13.934 tubular components, blastema components
NOTE Confidence: 0.94291633

01:34:14.395 --> 01:34:16.235 and stromal components are the
NOTE Confidence: 0.94291633

01:34:16.235 --> 01:34:18.260 rule. This is a blastema

NOTE Confidence: 0.94291633

01:34:18.400 --> 01:34:20.479 predominant component and this is

NOTE Confidence: 0.94291633

01:34:20.479 --> 01:34:21.460 a stromal predominant

NOTE Confidence: 0.9303192

01:34:21.840 --> 01:34:23.599 WIMS tumor. And sometimes you

NOTE Confidence: 0.9303192

01:34:23.599 --> 01:34:25.520 can have very clear muscle

NOTE Confidence: 0.9303192

01:34:25.520 --> 01:34:27.599 skeletal muscle differentiation that you

NOTE Confidence: 0.9303192

01:34:27.599 --> 01:34:29.219 can appreciate in these pictures

NOTE Confidence: 0.9303192

01:34:29.360 --> 01:34:30.420 from WIMS tumors.

NOTE Confidence: 0.90823835

01:34:30.785 --> 01:34:31.905 In the past, we used

NOTE Confidence: 0.90823835

01:34:31.905 --> 01:34:33.985 to call these teratoid variant

NOTE Confidence: 0.90823835

01:34:33.985 --> 01:34:35.185 of a Wilms tumor when

NOTE Confidence: 0.90823835

01:34:35.185 --> 01:34:36.785 there are components such as

NOTE Confidence: 0.90823835

01:34:36.785 --> 01:34:37.685 skeletal muscle,

NOTE Confidence: 0.8829753

01:34:38.705 --> 01:34:39.205 keratinizing

NOTE Confidence: 0.92407227

01:34:39.585 --> 01:34:41.845 squamous epithelium, and even osteoid

NOTE Confidence: 0.92407227

01:34:41.905 --> 01:34:42.405 formation.

NOTE Confidence: 0.8721191

01:34:43.790 --> 01:34:45.890 The war territory is not,
NOTE Confidence: 0.69055176

01:34:46.670 --> 01:34:47.810 recommended anymore.
NOTE Confidence: 0.9689209

01:34:48.430 --> 01:34:49.710 And after you make the
NOTE Confidence: 0.9689209

01:34:49.710 --> 01:34:51.810 diagnosis so what happens? Doctor,
NOTE Confidence: 0.28971353

01:34:54.030 --> 01:34:55.070 for Sanapan catch
NOTE Confidence: 0.97304285

01:34:55.630 --> 01:34:57.229 Pankar comes with you and
NOTE Confidence: 0.97304285

01:34:57.229 --> 01:34:58.210 tells you, Miguel,
NOTE Confidence: 0.82083565

01:34:58.955 --> 01:35:00.155 yesterday, there was an an
NOTE Confidence: 0.82083565

01:35:00.155 --> 01:35:01.215 infractomy specimen.
NOTE Confidence: 0.9313006

01:35:01.675 --> 01:35:02.715 Can you tell us what
NOTE Confidence: 0.9313006

01:35:02.715 --> 01:35:03.595 is it? Is it a
NOTE Confidence: 0.9313006

01:35:03.595 --> 01:35:04.715 wounds tumor? And then the
NOTE Confidence: 0.9313006

01:35:04.715 --> 01:35:06.015 first thing is to confirm.
NOTE Confidence: 0.9313006

01:35:06.075 --> 01:35:07.354 Yes. It's a wounds tumor,
NOTE Confidence: 0.9313006

01:35:07.354 --> 01:35:08.735 Farzana. Right? Good.
NOTE Confidence: 0.96203613

01:35:09.115 --> 01:35:09.935 Next question.

NOTE Confidence: 0.9904785
01:35:10.635 --> 01:35:11.695 What is the stage?
NOTE Confidence: 0.9691068
01:35:12.160 --> 01:35:13.760 Okay. And then is it
NOTE Confidence: 0.9691068
01:35:13.760 --> 01:35:15.840 favorable or unfavorable? And how
NOTE Confidence: 0.9691068
01:35:15.840 --> 01:35:17.280 do you distinguish that? Because
NOTE Confidence: 0.9691068
01:35:17.280 --> 01:35:18.880 that takes makes a difference
NOTE Confidence: 0.9691068
01:35:18.880 --> 01:35:20.080 in the treatment of the
NOTE Confidence: 0.9691068
01:35:20.080 --> 01:35:20.580 patient.
NOTE Confidence: 0.986883
01:35:21.040 --> 01:35:22.479 And then when I respond
NOTE Confidence: 0.986883
01:35:22.479 --> 01:35:24.320 these three questions, she goes
NOTE Confidence: 0.986883
01:35:24.320 --> 01:35:26.125 back to her office to
NOTE Confidence: 0.986883
01:35:26.125 --> 01:35:27.005 look at the patient and
NOTE Confidence: 0.986883
01:35:27.005 --> 01:35:27.825 start chemotherapy.
NOTE Confidence: 0.9995117
01:35:28.525 --> 01:35:29.425 Is that correct?
NOTE Confidence: 0.9824219
01:35:31.005 --> 01:35:31.505 However,
NOTE Confidence: 0.99316406
01:35:33.885 --> 01:35:35.585 favorable histology is
NOTE Confidence: 0.9839952

01:35:36.045 --> 01:35:37.585 identified with anaplastic
NOTE Confidence: 0.97555095

01:35:38.560 --> 01:35:41.520 mitosis or, hyperchromatic nucleomegaly. You
NOTE Confidence: 0.97555095

01:35:41.520 --> 01:35:42.400 have to have both of
NOTE Confidence: 0.97555095

01:35:42.400 --> 01:35:44.080 them for the diagnosis. I
NOTE Confidence: 0.97555095

01:35:44.080 --> 01:35:45.520 listed here the criteria. I'm
NOTE Confidence: 0.97555095

01:35:45.520 --> 01:35:46.880 not gonna go over them,
NOTE Confidence: 0.97555095

01:35:47.200 --> 01:35:47.939 in detail.
NOTE Confidence: 0.98128766

01:35:48.640 --> 01:35:50.580 This is another atypical mitosis
NOTE Confidence: 0.98128766

01:35:50.720 --> 01:35:52.755 and, hyperchromatic nucleomegaly
NOTE Confidence: 0.954575

01:35:53.055 --> 01:35:54.175 at least three times the
NOTE Confidence: 0.954575

01:35:54.175 --> 01:35:56.035 size of adjacent non anaplastic
NOTE Confidence: 0.954575

01:35:56.255 --> 01:35:56.755 nuclei.
NOTE Confidence: 0.96853065

01:35:57.375 --> 01:35:59.615 Immuno histochemistry is irrelevant in
NOTE Confidence: 0.96853065

01:35:59.615 --> 01:36:01.135 the diagnosis and study of
NOTE Confidence: 0.96853065

01:36:01.135 --> 01:36:02.895 WIMS tumor patients, but I
NOTE Confidence: 0.96853065

01:36:02.895 --> 01:36:04.950 listed here What is important

NOTE Confidence: 0.96853065

01:36:05.010 --> 01:36:06.310 is to add the molecular

NOTE Confidence: 0.96853065

01:36:06.370 --> 01:36:07.970 analysis for the patients because

NOTE Confidence: 0.96853065

01:36:07.970 --> 01:36:09.570 there are certain loss of

NOTE Confidence: 0.96853065

01:36:09.570 --> 01:36:10.070 heterozygosity

NOTE Confidence: 0.9980469

01:36:10.930 --> 01:36:12.710 and other chromosomal abnormalities

NOTE Confidence: 0.9750366

01:36:13.010 --> 01:36:14.390 that may determine treatment.

NOTE Confidence: 0.9962158

01:36:15.010 --> 01:36:15.970 And then you have to

NOTE Confidence: 0.9962158

01:36:15.970 --> 01:36:17.189 examine the residual

NOTE Confidence: 0.93762356

01:36:17.490 --> 01:36:19.775 kidney, the non tumor kidney.

NOTE Confidence: 0.93762356

01:36:19.835 --> 01:36:21.354 And in that, the important

NOTE Confidence: 0.93762356

01:36:21.354 --> 01:36:22.895 thing is to find nephrogenic

NOTE Confidence: 0.93762356

01:36:23.195 --> 01:36:24.475 rests because if there are

NOTE Confidence: 0.93762356

01:36:24.475 --> 01:36:26.554 nephrogenic rest, the possibility of

NOTE Confidence: 0.93762356

01:36:26.554 --> 01:36:27.215 a contralateral

NOTE Confidence: 0.8297555

01:36:27.915 --> 01:36:30.235 synchronous or metachronous once tumor

NOTE Confidence: 0.8297555

01:36:30.235 --> 01:36:30.895 is high.
NOTE Confidence: 0.982711

01:36:31.420 --> 01:36:32.700 And these are examples that
NOTE Confidence: 0.982711

01:36:32.700 --> 01:36:34.300 Bruce Beckwith gave me. These
NOTE Confidence: 0.982711

01:36:34.300 --> 01:36:36.220 are pictures from him where
NOTE Confidence: 0.982711

01:36:36.220 --> 01:36:37.360 this is the nephroblastomatosis.
NOTE Confidence: 0.9609187

01:36:38.620 --> 01:36:40.060 That word is only used
NOTE Confidence: 0.9609187

01:36:40.060 --> 01:36:41.340 for cases in which the
NOTE Confidence: 0.9609187

01:36:41.340 --> 01:36:42.640 entire renal parenchyma
NOTE Confidence: 0.96660155

01:36:43.020 --> 01:36:43.840 is nephroblastic
NOTE Confidence: 0.96477914

01:36:44.435 --> 01:36:46.035 and not a Wilms tumor
NOTE Confidence: 0.96477914

01:36:46.035 --> 01:36:47.395 directly. And this is an
NOTE Confidence: 0.96477914

01:36:47.395 --> 01:36:48.755 example of a Wilms tumor
NOTE Confidence: 0.96477914

01:36:48.755 --> 01:36:49.495 with numerous
NOTE Confidence: 0.7775608

01:36:49.875 --> 01:36:51.415 nephrogenic rest microscopically
NOTE Confidence: 0.92633057

01:36:51.955 --> 01:36:53.814 easy to identify. Okay.
NOTE Confidence: 0.98599523

01:36:54.435 --> 01:36:55.395 Let me give you an

NOTE Confidence: 0.98599523
01:36:55.395 --> 01:36:56.435 example of one of the
NOTE Confidence: 0.98599523
01:36:56.435 --> 01:36:57.795 last cases that I saw
NOTE Confidence: 0.98599523
01:36:57.795 --> 01:36:59.075 in Pittsburgh a couple of
NOTE Confidence: 0.98599523
01:36:59.075 --> 01:36:59.790 years ago.
NOTE Confidence: 0.9900545
01:37:00.430 --> 01:37:01.870 This tumor came from a
NOTE Confidence: 0.9900545
01:37:01.870 --> 01:37:02.910 two year old boy with
NOTE Confidence: 0.9900545
01:37:02.910 --> 01:37:04.030 a left renal mass. You
NOTE Confidence: 0.9900545
01:37:04.030 --> 01:37:05.090 can see the microscopic
NOTE Confidence: 0.8869629
01:37:05.390 --> 01:37:06.450 external appearance,
NOTE Confidence: 0.94659424
01:37:06.830 --> 01:37:08.450 the cross the surface,
NOTE Confidence: 0.95059484
01:37:09.550 --> 01:37:11.070 of the, section and the
NOTE Confidence: 0.95059484
01:37:11.070 --> 01:37:13.390 residual kidney is here. Everything
NOTE Confidence: 0.95059484
01:37:13.390 --> 01:37:13.890 fine?
NOTE Confidence: 0.9006754
01:37:14.695 --> 01:37:15.835 Blastema predominance,
NOTE Confidence: 0.95794517
01:37:16.455 --> 01:37:18.715 then there is, some perirenal
NOTE Confidence: 0.95794517

01:37:18.935 --> 01:37:20.055 fat, and then there is
NOTE Confidence: 0.95794517

01:37:20.055 --> 01:37:21.354 some residual kidney.
NOTE Confidence: 0.89457196

01:37:22.055 --> 01:37:23.495 So I gave the diagnosis.
NOTE Confidence: 0.89457196

01:37:23.495 --> 01:37:24.695 It's a woman's tumor. It's
NOTE Confidence: 0.89457196

01:37:24.695 --> 01:37:26.715 a favorable histology woman's tumor,
NOTE Confidence: 0.8779297

01:37:27.150 --> 01:37:28.350 and therefore favor,
NOTE Confidence: 0.9724571

01:37:28.830 --> 01:37:30.510 the chemotherapy is not as
NOTE Confidence: 0.9724571

01:37:30.510 --> 01:37:32.430 aggressive. And the stage is
NOTE Confidence: 0.9724571

01:37:32.430 --> 01:37:33.790 stage one because it hasn't
NOTE Confidence: 0.9724571

01:37:33.790 --> 01:37:35.010 broken through the capsule
NOTE Confidence: 0.9219055

01:37:35.390 --> 01:37:36.610 or the renal signs.
NOTE Confidence: 0.93967694

01:37:39.305 --> 01:37:41.145 And then you see, you,
NOTE Confidence: 0.93967694

01:37:41.145 --> 01:37:42.345 you, you, you show the
NOTE Confidence: 0.93967694

01:37:42.345 --> 01:37:43.085 three components
NOTE Confidence: 0.9399414

01:37:44.025 --> 01:37:44.985 and then you go to
NOTE Confidence: 0.9399414

01:37:44.985 --> 01:37:45.725 the renal

NOTE Confidence: 0.698999
01:37:46.345 --> 01:37:47.564 non tumoral parenchyma.
NOTE Confidence: 0.9986049
01:37:48.345 --> 01:37:49.705 And this is where things
NOTE Confidence: 0.9986049
01:37:49.705 --> 01:37:50.445 get interesting
NOTE Confidence: 0.51708984
01:37:51.145 --> 01:37:51.645 because
NOTE Confidence: 0.76708984
01:37:52.104 --> 01:37:53.705 diagnosing twins tumor is not
NOTE Confidence: 0.76708984
01:37:53.705 --> 01:37:54.205 difficult.
NOTE Confidence: 0.9729004
01:37:55.729 --> 01:37:57.350 But looking at this
NOTE Confidence: 0.8997803
01:37:58.050 --> 01:37:58.870 is difficult.
NOTE Confidence: 0.9764579
01:37:59.729 --> 01:38:01.010 Not looking at it, but
NOTE Confidence: 0.9764579
01:38:01.010 --> 01:38:01.750 in understanding
NOTE Confidence: 0.996148
01:38:02.130 --> 01:38:03.489 what you are seeing. What
NOTE Confidence: 0.996148
01:38:03.489 --> 01:38:04.949 you have here are
NOTE Confidence: 0.99768066
01:38:05.410 --> 01:38:07.189 tubular structures in the medulla
NOTE Confidence: 0.99768066
01:38:07.250 --> 01:38:08.150 that are separated
NOTE Confidence: 0.99576825
01:38:08.665 --> 01:38:09.485 by a relatively
NOTE Confidence: 0.81030273

01:38:09.864 --> 01:38:11.405 mixed soy type of
NOTE Confidence: 0.597229

01:38:11.784 --> 01:38:12.284 stroma
NOTE Confidence: 0.99072266

01:38:13.064 --> 01:38:14.985 that is not normal. This
NOTE Confidence: 0.99072266

01:38:14.985 --> 01:38:16.604 is not the normal morphology
NOTE Confidence: 0.98465985

01:38:17.145 --> 01:38:18.685 of the renal medulla.
NOTE Confidence: 0.9936035

01:38:19.064 --> 01:38:20.445 And when you see that,
NOTE Confidence: 0.9382551

01:38:21.460 --> 01:38:23.139 you have to diagnose it.
NOTE Confidence: 0.9382551

01:38:23.139 --> 01:38:24.579 So the diagnosis that I
NOTE Confidence: 0.9382551

01:38:24.579 --> 01:38:26.020 issued in this case is
NOTE Confidence: 0.9382551

01:38:26.020 --> 01:38:28.340 re is, written here. And
NOTE Confidence: 0.9382551

01:38:28.340 --> 01:38:30.360 then I put renal medullary
NOTE Confidence: 0.9382551

01:38:30.579 --> 01:38:32.979 dysplasia, consistent with Beckwith Wiedemann
NOTE Confidence: 0.9382551

01:38:32.979 --> 01:38:35.079 syndrome, see comment and references.
NOTE Confidence: 0.83266604

01:38:36.555 --> 01:38:37.995 When we discuss this case
NOTE Confidence: 0.83266604

01:38:37.995 --> 01:38:39.455 at tumor war in Pittsburgh,
NOTE Confidence: 0.92818934

01:38:40.955 --> 01:38:43.695 the, director of pediatric oncology,

NOTE Confidence: 0.92818934

01:38:43.835 --> 01:38:45.515 the clinical director of pediatric

NOTE Confidence: 0.92818934

01:38:45.515 --> 01:38:46.015 oncology

NOTE Confidence: 0.782959

01:38:46.315 --> 01:38:46.815 there,

NOTE Confidence: 0.8940582

01:38:47.595 --> 01:38:48.840 first I said this patient

NOTE Confidence: 0.8940582

01:38:48.840 --> 01:38:50.199 has Wilms tumor and this

NOTE Confidence: 0.8940582

01:38:50.199 --> 01:38:51.320 and this and then back

NOTE Confidence: 0.8940582

01:38:51.320 --> 01:38:53.000 with William and Cindy. And

NOTE Confidence: 0.8940582

01:38:53.000 --> 01:38:54.139 there was a silence.

NOTE Confidence: 0.9234794

01:38:55.320 --> 01:38:56.840 Very uncomfortable to be honest

NOTE Confidence: 0.9234794

01:38:56.840 --> 01:38:57.500 with you.

NOTE Confidence: 0.97089845

01:38:58.040 --> 01:38:59.900 And then he said, well,

NOTE Confidence: 0.8942871

01:39:00.520 --> 01:39:01.020 maybe.

NOTE Confidence: 0.9277649

01:39:03.514 --> 01:39:05.034 But, no, it's not a

NOTE Confidence: 0.9277649

01:39:05.034 --> 01:39:06.255 maybe. It is,

NOTE Confidence: 0.91778564

01:39:06.875 --> 01:39:09.375 Beckwith Wiedemann syndrome. I put

NOTE Confidence: 0.91778564

01:39:09.514 --> 01:39:09.915 this,
NOTE Confidence: 0.92800564

01:39:10.395 --> 01:39:11.995 comment in the report and
NOTE Confidence: 0.92800564

01:39:11.995 --> 01:39:13.195 I mentioned a previous,
NOTE Confidence: 0.92317706

01:39:14.074 --> 01:39:16.175 reference that Jorge Dotto,
NOTE Confidence: 0.9709636

01:39:16.530 --> 01:39:18.630 anybody here remembers Jorge Dotto?
NOTE Confidence: 0.97624326

01:39:19.250 --> 01:39:20.370 He wrote to me this
NOTE Confidence: 0.97624326

01:39:20.370 --> 01:39:21.410 morning because he saw the
NOTE Confidence: 0.97624326

01:39:21.410 --> 01:39:22.550 announcement in Twitter,
NOTE Confidence: 0.96828884

01:39:23.090 --> 01:39:24.370 of this conference, and he
NOTE Confidence: 0.96828884

01:39:24.370 --> 01:39:25.810 said, oh, Miguel. It's nice
NOTE Confidence: 0.96828884

01:39:25.810 --> 01:39:26.930 that you are going back
NOTE Confidence: 0.96828884

01:39:26.930 --> 01:39:28.229 to Yale. Well, I'm gonna
NOTE Confidence: 0.96828884

01:39:28.450 --> 01:39:29.810 show your name because he
NOTE Confidence: 0.96828884

01:39:29.810 --> 01:39:30.470 and I
NOTE Confidence: 0.9837036

01:39:31.125 --> 01:39:33.144 published a case that,
NOTE Confidence: 0.9988281

01:39:34.085 --> 01:39:35.784 was similar to this one.

NOTE Confidence: 0.92749023

01:39:36.085 --> 01:39:37.224 The case from Pittsburgh

NOTE Confidence: 0.99658203

01:39:37.844 --> 01:39:38.985 was published

NOTE Confidence: 0.9519531

01:39:39.925 --> 01:39:41.304 with this group of authors,

NOTE Confidence: 0.932684

01:39:41.604 --> 01:39:42.965 and one of them, Julia

NOTE Confidence: 0.932684

01:39:42.965 --> 01:39:44.724 Mead, is a pediatric, you

NOTE Confidence: 0.932684

01:39:44.724 --> 01:39:46.280 know, Julia Mead, Rosanna.

NOTE Confidence: 0.98063827

01:39:46.660 --> 01:39:48.340 She's a pediatric oncologist that

NOTE Confidence: 0.98063827

01:39:48.340 --> 01:39:50.200 is specialized in cancer predisposition

NOTE Confidence: 0.98063827

01:39:50.420 --> 01:39:51.220 syndromes. And,

NOTE Confidence: 0.97183084

01:39:51.620 --> 01:39:52.580 the following day of the

NOTE Confidence: 0.97183084

01:39:52.580 --> 01:39:53.620 tumor board, she comes to

NOTE Confidence: 0.97183084

01:39:53.620 --> 01:39:54.840 my office and she says,

NOTE Confidence: 0.97183084

01:39:55.060 --> 01:39:56.260 Miguel, can I cry with

NOTE Confidence: 0.97183084

01:39:56.260 --> 01:39:56.760 you?

NOTE Confidence: 0.99902344

01:39:57.300 --> 01:39:58.040 What happened?

NOTE Confidence: 0.99183875

01:39:59.115 --> 01:40:00.475 They you were right. They
NOTE Confidence: 0.99183875

01:40:00.475 --> 01:40:01.215 missed it.
NOTE Confidence: 0.94145066

01:40:01.515 --> 01:40:02.715 And the problem is that
NOTE Confidence: 0.94145066

01:40:02.715 --> 01:40:04.555 with, Beckwith Wiedemann syndrome is
NOTE Confidence: 0.94145066

01:40:04.555 --> 01:40:05.995 not an easy diagnosis to
NOTE Confidence: 0.94145066

01:40:05.995 --> 01:40:08.155 make even clinically, even to
NOTE Confidence: 0.94145066

01:40:08.155 --> 01:40:08.895 the specialist.
NOTE Confidence: 0.97744143

01:40:09.675 --> 01:40:11.615 I remember an autopsy here
NOTE Confidence: 0.9386776

01:40:12.800 --> 01:40:14.260 that one of our geneticists,
NOTE Confidence: 0.9386776

01:40:14.479 --> 01:40:16.900 a very highly respected geneticists
NOTE Confidence: 0.9386776

01:40:17.120 --> 01:40:18.880 here came to see. And
NOTE Confidence: 0.9386776

01:40:18.880 --> 01:40:19.540 when I
NOTE Confidence: 0.93722534

01:40:19.920 --> 01:40:20.880 just looked at it and
NOTE Confidence: 0.93722534

01:40:20.880 --> 01:40:21.680 I said, oh, it's a
NOTE Confidence: 0.93722534

01:40:21.680 --> 01:40:23.060 Beckwith Wiedemann syndrome.
NOTE Confidence: 0.9796956

01:40:23.360 --> 01:40:24.479 She looked at me like,

NOTE Confidence: 0.9796956
01:40:24.479 --> 01:40:24.979 what?
NOTE Confidence: 0.8157959
01:40:25.295 --> 01:40:26.114 And it was because
NOTE Confidence: 0.9930309
01:40:26.655 --> 01:40:27.935 it's not easy to diagnose
NOTE Confidence: 0.9930309
01:40:27.935 --> 01:40:29.215 it unless you think of
NOTE Confidence: 0.9930309
01:40:29.215 --> 01:40:29.715 it.
NOTE Confidence: 0.9770508
01:40:30.494 --> 01:40:30.994 So
NOTE Confidence: 0.95025635
01:40:31.775 --> 01:40:33.375 this patient that I show
NOTE Confidence: 0.95025635
01:40:33.375 --> 01:40:34.354 you was published
NOTE Confidence: 0.9290689
01:40:34.735 --> 01:40:37.074 and showing the medullary race,
NOTE Confidence: 0.98984104
01:40:37.540 --> 01:40:39.080 the medullary dysplasia,
NOTE Confidence: 0.9463542
01:40:39.460 --> 01:40:41.719 all the correlation with radiology,
NOTE Confidence: 0.92451984
01:40:42.100 --> 01:40:44.100 and then other examples in
NOTE Confidence: 0.92451984
01:40:44.100 --> 01:40:44.600 our,
NOTE Confidence: 0.90025264
01:40:46.260 --> 01:40:48.179 files were found with Beckwith
NOTE Confidence: 0.90025264
01:40:48.179 --> 01:40:50.085 Williammann syndrome and Wilf's tumor.
NOTE Confidence: 0.90025264

01:40:50.325 --> 01:40:51.844 What is Beckwith Wiedemann syndrome?

NOTE Confidence: 0.90025264

01:40:51.844 --> 01:40:53.205 Well, you know, it's a

NOTE Confidence: 0.90025264

01:40:53.205 --> 01:40:54.585 combination of overgrowth,

NOTE Confidence: 0.97073364

01:40:55.045 --> 01:40:57.604 affecting multiple organs described by

NOTE Confidence: 0.97073364

01:40:57.604 --> 01:40:58.905 Bruce Beckwith and,

NOTE Confidence: 0.9362793

01:40:59.285 --> 01:41:00.185 Hans Wiedemann,

NOTE Confidence: 0.93282646

01:41:00.885 --> 01:41:02.005 on a case report, by

NOTE Confidence: 0.93282646

01:41:02.005 --> 01:41:02.885 the way. And there are

NOTE Confidence: 0.93282646

01:41:02.885 --> 01:41:04.405 people that are very,

NOTE Confidence: 0.94869715

01:41:05.180 --> 01:41:06.300 upset when you say I'm

NOTE Confidence: 0.94869715

01:41:06.300 --> 01:41:07.100 going to do a case

NOTE Confidence: 0.94869715

01:41:07.100 --> 01:41:08.620 report and say what? You

NOTE Confidence: 0.94869715

01:41:08.620 --> 01:41:10.620 should that that's that's not

NOTE Confidence: 0.94869715

01:41:10.620 --> 01:41:11.439 good. Well,

NOTE Confidence: 0.89189756

01:41:11.820 --> 01:41:13.660 if Bruce of Witten or

NOTE Confidence: 0.89189756

01:41:13.660 --> 01:41:15.260 Witherman did not report that

NOTE Confidence: 0.89189756
01:41:15.260 --> 01:41:15.760 case,
NOTE Confidence: 0.9816963
01:41:16.380 --> 01:41:18.880 the field of overgrowth syndromes
NOTE Confidence: 0.9816963
01:41:19.020 --> 01:41:20.835 would not have started. Okay?
NOTE Confidence: 0.9816963
01:41:20.835 --> 01:41:21.695 So, yes,
NOTE Confidence: 0.9371338
01:41:22.155 --> 01:41:23.275 you may be wasting your
NOTE Confidence: 0.9371338
01:41:23.275 --> 01:41:25.034 time as I spoke this
NOTE Confidence: 0.9371338
01:41:25.034 --> 01:41:26.795 morning with doctor Ho how,
NOTE Confidence: 0.9371338
01:41:27.114 --> 01:41:27.614 go,
NOTE Confidence: 0.98949355
01:41:27.994 --> 01:41:29.675 on case reports, but you
NOTE Confidence: 0.98949355
01:41:29.675 --> 01:41:30.795 may not if you ask
NOTE Confidence: 0.98949355
01:41:30.795 --> 01:41:32.875 the interesting question. So don't
NOTE Confidence: 0.98949355
01:41:32.875 --> 01:41:34.715 don't don't get dissuaded to
NOTE Confidence: 0.98949355
01:41:34.715 --> 01:41:36.250 write cases if you think
NOTE Confidence: 0.98949355
01:41:36.250 --> 01:41:37.310 that they are interesting.
NOTE Confidence: 0.9650164
01:41:37.689 --> 01:41:38.989 These are all the genetic
NOTE Confidence: 0.9650164

01:41:39.050 --> 01:41:40.650 abnormalities. I'm not going to
NOTE Confidence: 0.9650164

01:41:40.650 --> 01:41:42.090 detail them because this is
NOTE Confidence: 0.9650164

01:41:42.090 --> 01:41:44.010 not the function, of this,
NOTE Confidence: 0.9650164

01:41:44.330 --> 01:41:45.530 conference. It's just to call
NOTE Confidence: 0.9650164

01:41:45.530 --> 01:41:46.889 your attention at why you
NOTE Confidence: 0.9650164

01:41:46.889 --> 01:41:48.409 need to look under the
NOTE Confidence: 0.9650164

01:41:48.409 --> 01:41:50.030 microscope or at the microscopic
NOTE Confidence: 0.9650164

01:41:50.250 --> 01:41:50.750 level.
NOTE Confidence: 0.8242409

01:41:52.385 --> 01:41:53.985 This is an example of
NOTE Confidence: 0.8242409

01:41:53.985 --> 01:41:55.345 a fetus with with the
NOTE Confidence: 0.8242409

01:41:55.345 --> 01:41:55.845 syndrome
NOTE Confidence: 0.9698731

01:41:57.185 --> 01:41:58.864 that we, carried on an
NOTE Confidence: 0.9698731

01:41:58.864 --> 01:42:01.285 autopsy here many years ago.
NOTE Confidence: 0.96998703

01:42:01.665 --> 01:42:02.864 You can see the exome
NOTE Confidence: 0.96998703

01:42:02.864 --> 01:42:04.485 follows. This is not umbilical
NOTE Confidence: 0.9874023

01:42:05.020 --> 01:42:06.460 hernia, but it's a huge

NOTE Confidence: 0.9874023
01:42:06.460 --> 01:42:09.199 umbilical hernia hernia that contains
NOTE Confidence: 0.94231623
01:42:09.500 --> 01:42:11.420 all the abdominal organs including
NOTE Confidence: 0.94231623
01:42:11.420 --> 01:42:12.699 the liver. And you see
NOTE Confidence: 0.94231623
01:42:12.699 --> 01:42:14.239 the the the two fetuses
NOTE Confidence: 0.94231623
01:42:14.300 --> 01:42:16.540 here with coarse features. They
NOTE Confidence: 0.94231623
01:42:16.540 --> 01:42:17.235 don't look
NOTE Confidence: 0.9996745
01:42:17.715 --> 01:42:19.074 nice. They are not cute
NOTE Confidence: 0.9996745
01:42:19.074 --> 01:42:19.574 babies.
NOTE Confidence: 0.9812012
01:42:20.114 --> 01:42:21.655 They are coarse because
NOTE Confidence: 0.99853516
01:42:22.114 --> 01:42:22.854 the overgrowth
NOTE Confidence: 0.98291016
01:42:23.235 --> 01:42:24.135 of the tissues,
NOTE Confidence: 0.9424642
01:42:24.435 --> 01:42:26.275 the soft tissues produces these
NOTE Confidence: 0.9424642
01:42:26.275 --> 01:42:26.775 creases
NOTE Confidence: 0.9958496
01:42:27.235 --> 01:42:27.975 and foldings
NOTE Confidence: 0.9982422
01:42:28.514 --> 01:42:30.295 that make them look not
NOTE Confidence: 0.99780273

01:42:31.000 --> 01:42:31.740 not cute.
NOTE Confidence: 0.9299164

01:42:32.280 --> 01:42:33.560 They have these creases in
NOTE Confidence: 0.9299164

01:42:33.560 --> 01:42:34.680 the ears as you can
NOTE Confidence: 0.9299164

01:42:34.680 --> 01:42:36.439 see here, these fissures and
NOTE Confidence: 0.9299164

01:42:36.439 --> 01:42:36.939 creases,
NOTE Confidence: 0.9670585

01:42:37.240 --> 01:42:38.840 and they obviously have different
NOTE Confidence: 0.9670585

01:42:38.840 --> 01:42:40.600 degrees of umbilical cord. And
NOTE Confidence: 0.9670585

01:42:40.600 --> 01:42:41.979 they can have hemihypertrophy
NOTE Confidence: 0.9962565

01:42:42.760 --> 01:42:43.500 or hypertrophy
NOTE Confidence: 0.99194336

01:42:43.880 --> 01:42:44.620 of something.
NOTE Confidence: 0.939767

01:42:45.315 --> 01:42:46.835 When I invited Bruce Beckwith
NOTE Confidence: 0.939767

01:42:46.835 --> 01:42:48.275 to give Grand Rounds right
NOTE Confidence: 0.939767

01:42:48.275 --> 01:42:49.895 here many years ago,
NOTE Confidence: 0.9329631

01:42:50.354 --> 01:42:52.515 we had lunch together and
NOTE Confidence: 0.9329631

01:42:52.515 --> 01:42:53.715 he told me, you know,
NOTE Confidence: 0.9329631

01:42:53.715 --> 01:42:54.455 the most,

NOTE Confidence: 0.99316406
01:42:56.435 --> 01:42:56.935 unforgettable
NOTE Confidence: 0.87320966
01:42:57.395 --> 01:42:58.950 case of he never said
NOTE Confidence: 0.87320966
01:42:58.950 --> 01:43:00.790 Beckwith Wiedemann syndrome. He said
NOTE Confidence: 0.87320966
01:43:00.790 --> 01:43:01.689 of BWS
NOTE Confidence: 0.98582107
01:43:03.030 --> 01:43:04.390 was a case that I
NOTE Confidence: 0.98582107
01:43:04.390 --> 01:43:05.510 saw on a man with
NOTE Confidence: 0.98582107
01:43:05.510 --> 01:43:06.570 a huge thumb.
NOTE Confidence: 0.91508216
01:43:07.430 --> 01:43:09.510 Okay? Because he doesn't hit
NOTE Confidence: 0.91508216
01:43:09.510 --> 01:43:11.189 the the normality doesn't hit
NOTE Confidence: 0.91508216
01:43:11.189 --> 01:43:12.790 all the organs at the
NOTE Confidence: 0.91508216
01:43:12.790 --> 01:43:13.610 same level.
NOTE Confidence: 0.9882702
01:43:15.875 --> 01:43:17.954 Okay? The fetal cortex in
NOTE Confidence: 0.9882702
01:43:17.954 --> 01:43:19.954 the adrenal medulla looks like
NOTE Confidence: 0.9882702
01:43:19.954 --> 01:43:21.715 this normally. This is the
NOTE Confidence: 0.9882702
01:43:21.715 --> 01:43:23.474 permanent cortex in the process
NOTE Confidence: 0.9882702

01:43:23.474 --> 01:43:24.755 of development. This is a
NOTE Confidence: 0.9882702

01:43:24.755 --> 01:43:26.354 fetus or a baby of
NOTE Confidence: 0.9882702

01:43:26.354 --> 01:43:27.795 less than six months of
NOTE Confidence: 0.9882702

01:43:27.795 --> 01:43:28.695 age postnatal.
NOTE Confidence: 0.9656738

01:43:29.370 --> 01:43:30.650 Okay? But it can be
NOTE Confidence: 0.9656738

01:43:30.650 --> 01:43:32.590 a newborn. And the pancreas
NOTE Confidence: 0.9411177

01:43:32.969 --> 01:43:34.570 may look like this. In
NOTE Confidence: 0.9411177

01:43:34.570 --> 01:43:36.330 Beckwith Wiedemann syndrome cases, you
NOTE Confidence: 0.9411177

01:43:36.330 --> 01:43:36.830 see
NOTE Confidence: 0.98095703

01:43:37.689 --> 01:43:38.189 nucleomegaly
NOTE Confidence: 0.99555665

01:43:38.729 --> 01:43:40.590 in the fetal adrenal cortex.
NOTE Confidence: 0.99780273

01:43:40.969 --> 01:43:42.875 That is a sign that
NOTE Confidence: 0.99780273

01:43:42.875 --> 01:43:44.415 is frequently associated
NOTE Confidence: 0.95327324

01:43:45.035 --> 01:43:47.515 to Beckwith Wiedemann syndrome. Not
NOTE Confidence: 0.95327324

01:43:47.515 --> 01:43:49.515 always. Sometimes it's just a
NOTE Confidence: 0.95327324

01:43:49.515 --> 01:43:50.795 sign of stress, but when

NOTE Confidence: 0.95327324

01:43:50.795 --> 01:43:51.995 it is when it is

NOTE Confidence: 0.95327324

01:43:51.995 --> 01:43:53.915 so diffuse, that's another clue

NOTE Confidence: 0.95327324

01:43:53.915 --> 01:43:55.614 that you should not miss.

NOTE Confidence: 0.95327324

01:43:55.880 --> 01:43:56.840 Because even if you are

NOTE Confidence: 0.95327324

01:43:56.840 --> 01:43:58.199 doing an autopsy or if

NOTE Confidence: 0.95327324

01:43:58.199 --> 01:43:59.880 you get a nephrectomy with

NOTE Confidence: 0.95327324

01:43:59.880 --> 01:44:01.239 an adrenal gland that looks

NOTE Confidence: 0.95327324

01:44:01.239 --> 01:44:02.360 like that, you need to

NOTE Confidence: 0.95327324

01:44:02.360 --> 01:44:02.860 call

NOTE Confidence: 0.9008586

01:44:03.800 --> 01:44:06.059 doctor Farzana and say,

NOTE Confidence: 0.8818534

01:44:07.320 --> 01:44:09.400 Beckwith Wiedemann syndrome. Please check

NOTE Confidence: 0.8818534

01:44:09.400 --> 01:44:10.219 for that.

NOTE Confidence: 0.6563721

01:44:12.564 --> 01:44:13.705 The CD of blastosis

NOTE Confidence: 0.96353513

01:44:14.085 --> 01:44:15.445 is the process of formation

NOTE Confidence: 0.96353513

01:44:15.445 --> 01:44:16.745 of the islets of Langerhans.

NOTE Confidence: 0.97068787

01:44:17.125 --> 01:44:18.805 And in Beckwith Wiedemann syndrome,

NOTE Confidence: 0.97068787

01:44:18.805 --> 01:44:19.925 you've had you have an

NOTE Confidence: 0.97068787

01:44:19.925 --> 01:44:22.085 increased number of islets of

NOTE Confidence: 0.97068787

01:44:22.085 --> 01:44:22.585 Langerhans.

NOTE Confidence: 0.953993

01:44:22.885 --> 01:44:24.370 This is a case that

NOTE Confidence: 0.953993

01:44:24.370 --> 01:44:26.130 I published with, Jorge Dotto.

NOTE Confidence: 0.953993

01:44:26.130 --> 01:44:26.930 And let me tell you

NOTE Confidence: 0.953993

01:44:26.930 --> 01:44:28.290 the story. When when this

NOTE Confidence: 0.953993

01:44:28.450 --> 01:44:29.330 you were not part of

NOTE Confidence: 0.953993

01:44:29.330 --> 01:44:30.530 the clinical team at that

NOTE Confidence: 0.953993

01:44:30.530 --> 01:44:31.970 time, I think, but they

NOTE Confidence: 0.953993

01:44:31.970 --> 01:44:33.890 came to the search part

NOTE Confidence: 0.953993

01:44:33.890 --> 01:44:35.330 area, and I was sitting

NOTE Confidence: 0.953993

01:44:35.330 --> 01:44:36.530 with Jorge looking at the

NOTE Confidence: 0.953993

01:44:36.530 --> 01:44:37.030 case.

NOTE Confidence: 0.959596

01:44:37.385 --> 01:44:39.064 And when, they asked me,

NOTE Confidence: 0.959596
01:44:39.064 --> 01:44:39.864 what is it? Is it
NOTE Confidence: 0.959596
01:44:39.864 --> 01:44:40.745 the Wilms? And I said,
NOTE Confidence: 0.959596
01:44:40.745 --> 01:44:41.625 yes. It is a Wilms
NOTE Confidence: 0.959596
01:44:41.625 --> 01:44:42.905 tumor. Is it favorable? Yes.
NOTE Confidence: 0.959596
01:44:42.905 --> 01:44:43.945 It is favorable. Thank you
NOTE Confidence: 0.959596
01:44:43.945 --> 01:44:45.005 very much. Okay.
NOTE Confidence: 0.98945314
01:44:45.305 --> 01:44:46.925 And then when I continue
NOTE Confidence: 0.99658203
01:44:47.385 --> 01:44:48.765 looking at the normal
NOTE Confidence: 0.9415283
01:44:49.145 --> 01:44:51.225 residual kidney, it is not
NOTE Confidence: 0.9415283
01:44:51.225 --> 01:44:51.725 normal.
NOTE Confidence: 0.94733536
01:44:52.265 --> 01:44:54.180 And I I asked Jorge
NOTE Confidence: 0.94733536
01:44:54.180 --> 01:44:55.220 and all the residents that
NOTE Confidence: 0.94733536
01:44:55.220 --> 01:44:56.739 were with me to walk
NOTE Confidence: 0.94733536
01:44:56.739 --> 01:44:58.100 to the bed of the
NOTE Confidence: 0.94733536
01:44:58.100 --> 01:44:58.600 patient.
NOTE Confidence: 0.961254

01:44:59.460 --> 01:45:01.620 Our our clinical colleagues looked

NOTE Confidence: 0.961254

01:45:01.620 --> 01:45:02.660 at us like, what are

NOTE Confidence: 0.961254

01:45:02.660 --> 01:45:03.860 these guys doing here on

NOTE Confidence: 0.961254

01:45:03.860 --> 01:45:05.320 the floor? These are pathologists.

NOTE Confidence: 0.961254

01:45:05.540 --> 01:45:07.060 Right? Well, I I asked

NOTE Confidence: 0.961254

01:45:07.060 --> 01:45:08.040 permission to

NOTE Confidence: 0.99365234

01:45:08.775 --> 01:45:10.075 check the external,

NOTE Confidence: 0.92944336

01:45:11.335 --> 01:45:12.155 the the clinical,

NOTE Confidence: 0.9291992

01:45:12.775 --> 01:45:13.895 appearance of the of the

NOTE Confidence: 0.9291992

01:45:13.895 --> 01:45:15.495 of the patient. It was

NOTE Confidence: 0.9291992

01:45:15.495 --> 01:45:16.635 a cute baby,

NOTE Confidence: 0.99609375

01:45:17.814 --> 01:45:19.115 about a year old

NOTE Confidence: 0.5263672

01:45:19.895 --> 01:45:20.395 and

NOTE Confidence: 0.8140082

01:45:21.679 --> 01:45:23.199 didn't have anything that looked

NOTE Confidence: 0.8140082

01:45:23.199 --> 01:45:24.560 like Bagel Guillain Man syndrome.

NOTE Confidence: 0.8140082

01:45:24.560 --> 01:45:25.460 No hemihypertrophy,

NOTE Confidence: 0.8987833
01:45:26.080 --> 01:45:28.020 no creases, nothing, not dysmorphology.
NOTE Confidence: 0.9777832
01:45:28.400 --> 01:45:29.699 And I was very disappointed.
NOTE Confidence: 0.95410156
01:45:30.479 --> 01:45:31.219 And I said,
NOTE Confidence: 0.9549037
01:45:31.920 --> 01:45:33.199 can't be. I sent a
NOTE Confidence: 0.9549037
01:45:33.199 --> 01:45:34.659 couple of pictures to Bruce
NOTE Confidence: 0.9549037
01:45:34.925 --> 01:45:35.965 and he calls me and
NOTE Confidence: 0.9549037
01:45:35.965 --> 01:45:38.205 he says, your patient has
NOTE Confidence: 0.9549037
01:45:38.205 --> 01:45:38.705 PWS
NOTE Confidence: 0.96203613
01:45:39.645 --> 01:45:40.685 whether the rest of his
NOTE Confidence: 0.96203613
01:45:40.685 --> 01:45:42.465 body knows it or not.
NOTE Confidence: 0.9890625
01:45:43.325 --> 01:45:45.425 And they prove it genetically.
NOTE Confidence: 0.9980469
01:45:46.285 --> 01:45:46.785 So
NOTE Confidence: 0.9511719
01:45:47.330 --> 01:45:48.610 this is a type of
NOTE Confidence: 0.9511719
01:45:48.610 --> 01:45:49.650 malformation or,
NOTE Confidence: 0.99902344
01:45:50.370 --> 01:45:50.870 abnormal
NOTE Confidence: 0.8924484

01:45:51.250 --> 01:45:53.090 development of the renal medulla
NOTE Confidence: 0.8924484

01:45:53.090 --> 01:45:54.210 that you can appreciate in
NOTE Confidence: 0.8924484

01:45:54.210 --> 01:45:55.970 Beckwith Wiedemann syndrome with this
NOTE Confidence: 0.8924484

01:45:55.970 --> 01:45:56.470 mixedoid
NOTE Confidence: 0.96269757

01:45:56.930 --> 01:45:58.530 stroma around the tubules and
NOTE Confidence: 0.96269757

01:45:58.530 --> 01:46:00.310 the separation of the tubules.
NOTE Confidence: 0.96269757

01:46:00.610 --> 01:46:02.205 Please pay attention to that
NOTE Confidence: 0.96269757

01:46:02.285 --> 01:46:03.485 in those cases. You can
NOTE Confidence: 0.96269757

01:46:03.485 --> 01:46:05.165 also see nephrogenic rest. This
NOTE Confidence: 0.96269757

01:46:05.165 --> 01:46:06.685 is another example with a
NOTE Confidence: 0.96269757

01:46:06.685 --> 01:46:08.445 medullary ray, and these are
NOTE Confidence: 0.96269757

01:46:08.445 --> 01:46:10.625 the usual molecular subtypes
NOTE Confidence: 0.87698644

01:46:11.085 --> 01:46:12.765 of Beckwith Wiedemann syndrome. This
NOTE Confidence: 0.87698644

01:46:12.765 --> 01:46:14.604 is taken from the PETES
NOTE Confidence: 0.87698644

01:46:14.604 --> 01:46:15.005 five,
NOTE Confidence: 0.84375

01:46:15.725 --> 01:46:17.104 edition of the WHO

NOTE Confidence: 0.98828125
01:46:18.570 --> 01:46:19.869 classification of tumors,
NOTE Confidence: 0.9700753
01:46:20.330 --> 01:46:21.929 and we put these pictures
NOTE Confidence: 0.9700753
01:46:21.929 --> 01:46:22.590 in that,
NOTE Confidence: 1
01:46:23.130 --> 01:46:23.630 volume.
NOTE Confidence: 0.96362305
01:46:24.489 --> 01:46:26.030 Other tumors may also,
NOTE Confidence: 0.94790417
01:46:27.369 --> 01:46:29.050 appear in patients with Beckwith
NOTE Confidence: 0.94790417
01:46:29.050 --> 01:46:30.729 Wiedemann syndrome, of course. This
NOTE Confidence: 0.94790417
01:46:30.729 --> 01:46:31.550 is a case,
NOTE Confidence: 0.892334
01:46:32.285 --> 01:46:32.765 that,
NOTE Confidence: 0.9814453
01:46:33.165 --> 01:46:34.625 had a, mesenchymal
NOTE Confidence: 0.9845378
01:46:34.925 --> 01:46:35.425 hamartoma
NOTE Confidence: 0.9822998
01:46:36.045 --> 01:46:36.845 associated with,
NOTE Confidence: 0.94213593
01:46:37.405 --> 01:46:38.925 Beckwith Guiedemann syndrome. By the
NOTE Confidence: 0.94213593
01:46:38.925 --> 01:46:40.765 way, the alpha fetoprotein in
NOTE Confidence: 0.94213593
01:46:40.765 --> 01:46:42.225 this patient was a million
NOTE Confidence: 0.7234294

01:46:42.685 --> 01:46:43.665 above a million,
NOTE Confidence: 0.9383057

01:46:44.285 --> 01:46:45.405 and it was not a
NOTE Confidence: 0.9383057

01:46:45.405 --> 01:46:45.905 hepatoblastoma,
NOTE Confidence: 0.95480347

01:46:46.365 --> 01:46:48.180 which what would have been
NOTE Confidence: 0.95480347

01:46:48.180 --> 01:46:49.479 the first suspicion.
NOTE Confidence: 0.9871419

01:46:50.180 --> 01:46:51.460 Okay? But it's not a
NOTE Confidence: 0.9871419

01:46:51.460 --> 01:46:51.960 hepatoblastoma.
NOTE Confidence: 0.97369385

01:46:52.340 --> 01:46:53.300 I remember we sent it
NOTE Confidence: 0.97369385

01:46:53.300 --> 01:46:54.520 to Milton Feingold
NOTE Confidence: 0.88578033

01:46:54.979 --> 01:46:56.900 at Texas Children's Hospital and
NOTE Confidence: 0.88578033

01:46:56.900 --> 01:46:57.780 he called me. He said,
NOTE Confidence: 0.88578033

01:46:57.780 --> 01:46:59.220 you're right, Miguel. He said,
NOTE Confidence: 0.88578033

01:46:59.220 --> 01:46:59.720 mesenchymal
NOTE Confidence: 0.97055286

01:47:00.420 --> 01:47:01.620 glaucoma. So we need to
NOTE Confidence: 0.97055286

01:47:01.620 --> 01:47:03.165 pay attention because the morphology
NOTE Confidence: 0.97055286

01:47:03.465 --> 01:47:04.364 of these lesions

NOTE Confidence: 0.9970703
01:47:04.824 --> 01:47:05.324 may
NOTE Confidence: 0.99550086
01:47:05.864 --> 01:47:07.225 give you the clue and
NOTE Confidence: 0.99550086
01:47:07.225 --> 01:47:07.885 the diagnosis,
NOTE Confidence: 0.95084053
01:47:08.985 --> 01:47:10.905 of Beckwith Wiedemann syndrome. Let's
NOTE Confidence: 0.95084053
01:47:10.905 --> 01:47:11.885 change organs.
NOTE Confidence: 0.9694824
01:47:14.600 --> 01:47:16.200 I'm sure that you all
NOTE Confidence: 0.9694824
01:47:16.200 --> 01:47:18.520 have heard, used, and applied
NOTE Confidence: 0.9694824
01:47:18.520 --> 01:47:19.180 the term
NOTE Confidence: 0.90722656
01:47:19.640 --> 01:47:20.140 CPAM
NOTE Confidence: 0.97566736
01:47:20.600 --> 01:47:21.420 or CCAM.
NOTE Confidence: 0.97859704
01:47:22.040 --> 01:47:23.340 Right? Cystic,
NOTE Confidence: 0.9105469
01:47:25.080 --> 01:47:25.580 adenomatoid
NOTE Confidence: 0.9783325
01:47:25.960 --> 01:47:27.420 malformation of the lung
NOTE Confidence: 0.9519043
01:47:27.745 --> 01:47:28.885 or cystic pulmonary,
NOTE Confidence: 0.9960327
01:47:29.985 --> 01:47:31.445 malformation of the lung.
NOTE Confidence: 0.9980469

01:47:31.985 --> 01:47:32.485 Well,
NOTE Confidence: 0.99726564
01:47:33.745 --> 01:47:35.125 please don't use it anymore.
NOTE Confidence: 0.9995117
01:47:36.145 --> 01:47:36.645 It's
NOTE Confidence: 0.97190636
01:47:37.104 --> 01:47:39.045 changing. The nomenclature is being
NOTE Confidence: 0.97190636
01:47:39.104 --> 01:47:40.545 updated. I haven't made that
NOTE Confidence: 0.97190636
01:47:40.545 --> 01:47:42.645 diagnosis in about fourteen years.
NOTE Confidence: 0.97190636
01:47:42.920 --> 01:47:44.300 There are people that write
NOTE Confidence: 0.97190636
01:47:44.599 --> 01:47:45.979 and that are very nice
NOTE Confidence: 0.97190636
01:47:46.280 --> 01:47:47.960 and trying to please people
NOTE Confidence: 0.97190636
01:47:47.960 --> 01:47:49.960 that have described this. Among
NOTE Confidence: 0.97190636
01:47:49.960 --> 01:47:51.800 them, a good friend, Doctor.
NOTE Confidence: 0.97190636
01:47:51.800 --> 01:47:52.619 Tom Stalker,
NOTE Confidence: 0.94538224
01:47:52.920 --> 01:47:54.780 a retired professor who made
NOTE Confidence: 0.94538224
01:47:54.840 --> 01:47:55.659 all these
NOTE Confidence: 0.99902344
01:47:55.960 --> 01:47:56.460 very
NOTE Confidence: 0.9001465
01:47:57.000 --> 01:47:57.500 intuitive,

NOTE Confidence: 0.8598633
01:47:58.564 --> 01:47:59.064 beautiful,
NOTE Confidence: 1
01:47:59.764 --> 01:48:00.264 conceptual
NOTE Confidence: 0.9667969
01:48:00.885 --> 01:48:01.385 understanding
NOTE Confidence: 0.964386
01:48:02.005 --> 01:48:03.604 of the development of the
NOTE Confidence: 0.964386
01:48:03.604 --> 01:48:05.125 lung and the cystic lesions.
NOTE Confidence: 0.964386
01:48:05.125 --> 01:48:06.564 The problem is that however
NOTE Confidence: 0.964386
01:48:06.564 --> 01:48:08.885 beautiful this was in types
NOTE Confidence: 0.964386
01:48:08.885 --> 01:48:10.665 zero to type four,
NOTE Confidence: 0.9501953
01:48:11.364 --> 01:48:12.264 it's wrong.
NOTE Confidence: 0.96069336
01:48:13.600 --> 01:48:15.280 And, therefore, we need to
NOTE Confidence: 0.96069336
01:48:15.280 --> 01:48:15.780 move
NOTE Confidence: 0.9992676
01:48:16.320 --> 01:48:17.920 to use a more current
NOTE Confidence: 0.9992676
01:48:17.920 --> 01:48:18.420 classification.
NOTE Confidence: 0.90625674
01:48:19.360 --> 01:48:21.620 There are classically three types,
NOTE Confidence: 0.90625674
01:48:21.680 --> 01:48:23.780 large c's, medium sized c's,
NOTE Confidence: 0.90625674

01:48:23.840 --> 01:48:25.440 small c's, and then he
NOTE Confidence: 0.90625674

01:48:25.440 --> 01:48:27.040 said, well, this occurred because
NOTE Confidence: 0.90625674

01:48:27.040 --> 01:48:28.455 they develop in the
NOTE Confidence: 0.92041016

01:48:28.935 --> 01:48:29.435 bronchi,
NOTE Confidence: 0.88533527

01:48:29.895 --> 01:48:32.215 terminal bronchioles or alveoli. Well,
NOTE Confidence: 0.88533527

01:48:32.215 --> 01:48:32.615 no.
NOTE Confidence: 0.94609725

01:48:33.175 --> 01:48:34.615 She added two more types,
NOTE Confidence: 0.94609725

01:48:34.615 --> 01:48:35.915 etcetera. No.
NOTE Confidence: 0.957798

01:48:36.615 --> 01:48:37.975 There are other things that
NOTE Confidence: 0.957798

01:48:37.975 --> 01:48:39.735 look a little bit different
NOTE Confidence: 0.957798

01:48:39.735 --> 01:48:40.535 but are part of the
NOTE Confidence: 0.957798

01:48:40.535 --> 01:48:42.795 same spectrum. Congenital lower emphysema
NOTE Confidence: 0.957798

01:48:42.935 --> 01:48:44.430 is one of them. This
NOTE Confidence: 0.957798

01:48:44.430 --> 01:48:46.670 dilatation of the lung with
NOTE Confidence: 0.957798

01:48:46.670 --> 01:48:47.250 a hyper,
NOTE Confidence: 0.7819824

01:48:49.790 --> 01:48:50.290 distension

NOTE Confidence: 0.9575417
01:48:50.830 --> 01:48:51.870 and you can see the
NOTE Confidence: 0.9575417
01:48:51.870 --> 01:48:53.630 microscopic appearance of this long
NOTE Confidence: 0.9575417
01:48:53.630 --> 01:48:54.130 parenchyma
NOTE Confidence: 0.9251758
01:48:54.590 --> 01:48:56.350 and sometimes you can see
NOTE Confidence: 0.9251758
01:48:56.350 --> 01:48:58.130 like this under the diaphragm.
NOTE Confidence: 0.9485779
01:48:59.205 --> 01:49:00.005 There is a piece of
NOTE Confidence: 0.9485779
01:49:00.005 --> 01:49:01.205 lung that is very solid.
NOTE Confidence: 0.9485779
01:49:01.205 --> 01:49:02.324 This is called the Rocky
NOTE Confidence: 0.9485779
01:49:02.324 --> 01:49:02.824 Tanski
NOTE Confidence: 0.9375
01:49:03.284 --> 01:49:03.784 lobe
NOTE Confidence: 0.9621887
01:49:04.165 --> 01:49:05.685 of the lung, which is
NOTE Confidence: 0.9621887
01:49:05.685 --> 01:49:06.804 part of the,
NOTE Confidence: 0.9996745
01:49:07.445 --> 01:49:08.665 spectrum of disorders
NOTE Confidence: 0.99121094
01:49:09.125 --> 01:49:09.945 that includes
NOTE Confidence: 0.93880206
01:49:10.405 --> 01:49:10.905 sequestrations,
NOTE Confidence: 0.9382054

01:49:12.300 --> 01:49:13.900 so called or formerly called
NOTE Confidence: 0.9382054

01:49:13.900 --> 01:49:16.080 CPAMs, and congenital lower emphysema.
NOTE Confidence: 0.9382054

01:49:16.300 --> 01:49:17.580 All of those lesions come
NOTE Confidence: 0.9382054

01:49:17.580 --> 01:49:19.040 to us pediatric pathologies
NOTE Confidence: 0.9693447

01:49:19.580 --> 01:49:21.580 very frequently. I don't know,
NOTE Confidence: 0.9693447

01:49:21.580 --> 01:49:23.740 Rafaela, you must receive about
NOTE Confidence: 0.9693447

01:49:23.740 --> 01:49:24.800 one every month
NOTE Confidence: 0.96895105

01:49:25.455 --> 01:49:27.935 probably or more. Right? Because
NOTE Confidence: 0.96895105

01:49:27.935 --> 01:49:28.974 that's more or less what
NOTE Confidence: 0.96895105

01:49:28.974 --> 01:49:30.175 I remember and they look
NOTE Confidence: 0.96895105

01:49:30.175 --> 01:49:30.835 like this.
NOTE Confidence: 0.99853516

01:49:31.455 --> 01:49:32.195 And we
NOTE Confidence: 0.9968959

01:49:32.574 --> 01:49:34.014 usually don't have a problem
NOTE Confidence: 0.9968959

01:49:34.014 --> 01:49:35.235 diagnosing this.
NOTE Confidence: 0.9995117

01:49:35.695 --> 01:49:36.435 The problem
NOTE Confidence: 0.9456692

01:49:37.135 --> 01:49:38.335 well, sometimes this is a

NOTE Confidence: 0.9456692

01:49:38.335 --> 01:49:39.580 Rocky Tanske lob. See, this

NOTE Confidence: 0.9456692

01:49:39.580 --> 01:49:40.540 is a picture I took

NOTE Confidence: 0.9456692

01:49:40.540 --> 01:49:42.300 in the eighties in Mexico

NOTE Confidence: 0.9456692

01:49:42.300 --> 01:49:43.900 City where you see the

NOTE Confidence: 0.9456692

01:49:43.900 --> 01:49:45.420 two lungs and then this

NOTE Confidence: 0.9456692

01:49:45.420 --> 01:49:47.260 lobe with a pedicle and

NOTE Confidence: 0.9456692

01:49:47.260 --> 01:49:48.880 vessels. This is a typical

NOTE Confidence: 0.9456692

01:49:49.020 --> 01:49:50.780 sequestration, and we published it

NOTE Confidence: 0.9456692

01:49:50.780 --> 01:49:52.264 with Mark Keller. I don't

NOTE Confidence: 0.9456692

01:49:52.264 --> 01:49:54.045 know if anybody here remembers

NOTE Confidence: 0.9456692

01:49:54.104 --> 01:49:55.465 Mark Keller. He now lives

NOTE Confidence: 0.9456692

01:49:55.465 --> 01:49:57.064 in Miami, close to Miami,

NOTE Confidence: 0.9456692

01:49:57.064 --> 01:49:57.945 and he was the chief

NOTE Confidence: 0.9456692

01:49:57.945 --> 01:49:59.304 of radiology here, and we

NOTE Confidence: 0.9456692

01:49:59.304 --> 01:50:01.224 just had dinner together last

NOTE Confidence: 0.9456692

01:50:01.224 --> 01:50:03.405 week. It's a wonderful colleague.
NOTE Confidence: 0.98187256

01:50:03.864 --> 01:50:05.224 Well, the problem is that
NOTE Confidence: 0.98187256

01:50:05.224 --> 01:50:06.665 many years ago, the group
NOTE Confidence: 0.98187256

01:50:06.665 --> 01:50:07.324 at Harvard,
NOTE Confidence: 0.93890566

01:50:08.000 --> 01:50:09.760 Boston Children's, the h word
NOTE Confidence: 0.93890566

01:50:09.840 --> 01:50:11.120 I'm sorry, John. I remember
NOTE Confidence: 0.93890566

01:50:11.120 --> 01:50:12.239 that the h word is,
NOTE Confidence: 0.93890566

01:50:12.479 --> 01:50:14.239 is is problematic here, that
NOTE Confidence: 0.93890566

01:50:14.239 --> 01:50:15.439 evil place of the north
NOTE Confidence: 0.93890566

01:50:15.439 --> 01:50:16.640 as you used to call
NOTE Confidence: 0.93890566

01:50:16.640 --> 01:50:17.840 it, John. But,
NOTE Confidence: 0.8732503

01:50:18.719 --> 01:50:20.979 when the Boston Children's describe
NOTE Confidence: 0.8732503

01:50:21.120 --> 01:50:21.620 this
NOTE Confidence: 0.9019593

01:50:22.215 --> 01:50:24.795 paper, he's they said bronchial
NOTE Confidence: 0.9019593

01:50:24.935 --> 01:50:26.854 atresia is common to extra
NOTE Confidence: 0.9019593

01:50:26.854 --> 01:50:28.955 lower sequestration, intra lower sequestration,

NOTE Confidence: 0.9019593
01:50:29.095 --> 01:50:30.935 and c cam and lower
NOTE Confidence: 0.9019593
01:50:30.935 --> 01:50:31.435 emphysema.
NOTE Confidence: 0.90752774
01:50:32.055 --> 01:50:33.415 So when you obstruct the
NOTE Confidence: 0.90752774
01:50:33.415 --> 01:50:35.995 bronchus in a developing kid,
NOTE Confidence: 0.97177356
01:50:36.340 --> 01:50:37.780 that is what happens. Any
NOTE Confidence: 0.97177356
01:50:37.780 --> 01:50:38.900 of those lesions that I
NOTE Confidence: 0.97177356
01:50:38.900 --> 01:50:40.520 mentioned happens. Okay.
NOTE Confidence: 0.9988356
01:50:41.060 --> 01:50:42.180 And there are many papers
NOTE Confidence: 0.9988356
01:50:42.180 --> 01:50:44.260 supporting that. The problem is
NOTE Confidence: 0.9988356
01:50:44.260 --> 01:50:45.160 when you receive
NOTE Confidence: 0.9876302
01:50:45.620 --> 01:50:46.680 something like this.
NOTE Confidence: 0.94426906
01:50:47.300 --> 01:50:48.580 You receive something like this
NOTE Confidence: 0.94426906
01:50:48.580 --> 01:50:49.540 and then you have the
NOTE Confidence: 0.94426906
01:50:49.540 --> 01:50:50.660 cyst here and you say,
NOTE Confidence: 0.94426906
01:50:50.660 --> 01:50:52.155 well, this may be a
NOTE Confidence: 0.94426906

01:50:52.235 --> 01:50:54.314 bronchial atresia deformation sequence or
NOTE Confidence: 0.94426906

01:50:54.314 --> 01:50:55.375 so called CPAM
NOTE Confidence: 0.91240233

01:50:55.835 --> 01:50:57.215 and you see this and
NOTE Confidence: 0.9864163

01:50:58.314 --> 01:50:59.675 you have to consider that
NOTE Confidence: 0.9864163

01:50:59.675 --> 01:51:00.954 this may be a pleural
NOTE Confidence: 0.9864163

01:51:00.954 --> 01:51:02.094 pulmonary blastoma.
NOTE Confidence: 0.96984863

01:51:03.034 --> 01:51:04.895 This is a very aggressive
NOTE Confidence: 0.96984863

01:51:04.954 --> 01:51:06.175 in general tumor
NOTE Confidence: 0.99902344

01:51:06.530 --> 01:51:07.969 that occurs by
NOTE Confidence: 0.91101074

01:51:08.330 --> 01:51:10.450 with mutations of Dicer one.
NOTE Confidence: 0.91101074

01:51:10.450 --> 01:51:11.270 I'm sorry.
NOTE Confidence: 0.9101737

01:51:13.170 --> 01:51:15.810 Dicer one. And it occurs
NOTE Confidence: 0.9101737

01:51:15.810 --> 01:51:16.469 in three
NOTE Confidence: 0.945757

01:51:16.770 --> 01:51:18.689 subtypes, one, two, and three,
NOTE Confidence: 0.945757

01:51:18.689 --> 01:51:20.295 and one is called one,
NOTE Confidence: 0.945757

01:51:20.515 --> 01:51:21.655 type one regressing.

NOTE Confidence: 0.90372723

01:51:22.195 --> 01:51:23.395 It occurs in the lungs

NOTE Confidence: 0.90372723

01:51:23.395 --> 01:51:24.595 and derives from the long

NOTE Confidence: 0.90372723

01:51:24.595 --> 01:51:26.115 missing kind. But now we

NOTE Confidence: 0.90372723

01:51:26.115 --> 01:51:27.555 are learning that dicer one

NOTE Confidence: 0.90372723

01:51:27.555 --> 01:51:29.095 mutations can produce tumors

NOTE Confidence: 0.99820966

01:51:29.555 --> 01:51:31.335 anywhere and everywhere.

NOTE Confidence: 0.99746096

01:51:31.795 --> 01:51:33.335 The first ones were described

NOTE Confidence: 0.9320679

01:51:33.635 --> 01:51:35.270 in the lung. Okay?

NOTE Confidence: 0.92941624

01:51:36.530 --> 01:51:38.050 They were described by two

NOTE Confidence: 0.92941624

01:51:38.050 --> 01:51:39.969 good friends, Pepper Dehner and

NOTE Confidence: 0.92941624

01:51:39.969 --> 01:51:41.030 Carlos Manibal,

NOTE Confidence: 0.9941101

01:51:42.050 --> 01:51:43.329 who taught me how to

NOTE Confidence: 0.9941101

01:51:43.329 --> 01:51:44.610 do an autopsy for the

NOTE Confidence: 0.9941101

01:51:44.610 --> 01:51:46.689 first time in nineteen eighty

NOTE Confidence: 0.9941101

01:51:46.689 --> 01:51:47.189 six

NOTE Confidence: 0.9839274

01:51:47.875 --> 01:51:49.074 or eighty four. I don't
NOTE Confidence: 0.9839274

01:51:49.074 --> 01:51:49.574 remember.
NOTE Confidence: 0.9926758

01:51:50.275 --> 01:51:51.655 These two guys in Minneapolis,
NOTE Confidence: 0.9623108

01:51:52.354 --> 01:51:53.474 Pepperdiner is one of the
NOTE Confidence: 0.9623108

01:51:53.474 --> 01:51:55.554 most prominent pediatric pathologists in
NOTE Confidence: 0.9623108

01:51:55.554 --> 01:51:57.395 history. And Carlos is a
NOTE Confidence: 0.9623108

01:51:57.395 --> 01:51:59.155 pediatric pathologist at heart, but
NOTE Confidence: 0.9623108

01:51:59.155 --> 01:52:00.375 he's a general pathologist.
NOTE Confidence: 0.92323524

01:52:01.074 --> 01:52:03.030 And they describe this pleural
NOTE Confidence: 0.92323524

01:52:03.030 --> 01:52:04.090 pulmonary blastoma
NOTE Confidence: 0.9992676

01:52:04.550 --> 01:52:06.489 in nineteen eighty eight.
NOTE Confidence: 0.96533203

01:52:07.030 --> 01:52:07.530 Well,
NOTE Confidence: 0.98205566

01:52:08.070 --> 01:52:08.889 after that,
NOTE Confidence: 0.94677734

01:52:09.670 --> 01:52:11.210 Ashley Hill, Jason
NOTE Confidence: 0.4051107

01:52:11.670 --> 01:52:12.809 Jason Jarsimboski,
NOTE Confidence: 0.8623454

01:52:13.189 --> 01:52:15.189 and Pepper Dehner describe the

NOTE Confidence: 0.8623454
01:52:15.189 --> 01:52:17.210 mutations on these lesions.
NOTE Confidence: 0.9555664
01:52:17.755 --> 01:52:18.415 And then,
NOTE Confidence: 0.9458008
01:52:19.115 --> 01:52:21.195 identify the major genetic factor
NOTE Confidence: 0.9458008
01:52:21.195 --> 01:52:22.575 which is Dicer one
NOTE Confidence: 0.9468689
01:52:23.115 --> 01:52:24.715 mutations that lead to the
NOTE Confidence: 0.9468689
01:52:24.715 --> 01:52:25.935 Dicer one syndrome.
NOTE Confidence: 0.8222656
01:52:26.555 --> 01:52:27.055 These,
NOTE Confidence: 0.8666992
01:52:27.755 --> 01:52:28.235 are,
NOTE Confidence: 0.9565492
01:52:28.875 --> 01:52:30.780 mutations that affect a key
NOTE Confidence: 0.9565492
01:52:30.780 --> 01:52:32.719 enzyme required to keep precursors
NOTE Confidence: 0.9565492
01:52:32.860 --> 01:52:34.239 of micro RNAs
NOTE Confidence: 0.95080566
01:52:34.939 --> 01:52:37.199 into their mature active forms.
NOTE Confidence: 0.95080566
01:52:37.420 --> 01:52:38.719 And when they are deleted,
NOTE Confidence: 0.93269855
01:52:39.500 --> 01:52:41.040 they are, very,
NOTE Confidence: 0.9852702
01:52:41.979 --> 01:52:43.199 severe in effects.
NOTE Confidence: 0.8782959

01:52:44.025 --> 01:52:45.245 The, germline,
NOTE Confidence: 0.9980469
01:52:45.705 --> 01:52:46.205 pathogenic
NOTE Confidence: 0.824242
01:52:46.665 --> 01:52:48.344 Dicer one variants, they find
NOTE Confidence: 0.824242
01:52:48.344 --> 01:52:49.085 these syndromes
NOTE Confidence: 0.94588214
01:52:49.465 --> 01:52:51.465 are always loss of function
NOTE Confidence: 0.94588214
01:52:51.465 --> 01:52:51.965 type
NOTE Confidence: 0.8980487
01:52:52.425 --> 01:52:54.025 and, like many other tumor
NOTE Confidence: 0.8980487
01:52:54.025 --> 01:52:56.745 suppressor genes, germline, a loss
NOTE Confidence: 0.8980487
01:52:56.745 --> 01:52:58.344 of function mutations in Dicer
NOTE Confidence: 0.8980487
01:52:58.344 --> 01:52:59.324 one create tumor
NOTE Confidence: 0.9970703
01:52:59.780 --> 01:53:00.280 susceptibility
NOTE Confidence: 0.9549484
01:53:00.660 --> 01:53:02.500 but appear to be insufficient
NOTE Confidence: 0.9549484
01:53:02.500 --> 01:53:03.800 to initiate tumorigenesis.
NOTE Confidence: 0.95462036
01:53:04.420 --> 01:53:06.340 There are other changes required
NOTE Confidence: 0.95462036
01:53:06.340 --> 01:53:07.479 to develop into,
NOTE Confidence: 0.9884033
01:53:08.020 --> 01:53:08.520 tumorigenesis.

NOTE Confidence: 0.9986328

01:53:09.460 --> 01:53:11.160 Many different organs are affected.

NOTE Confidence: 0.96698576

01:53:11.715 --> 01:53:12.915 The first one described was

NOTE Confidence: 0.96698576

01:53:12.915 --> 01:53:14.915 pluripulmonary blastoma, but we now

NOTE Confidence: 0.96698576

01:53:14.915 --> 01:53:16.375 know that cystic nephroma

NOTE Confidence: 0.90234715

01:53:16.675 --> 01:53:18.195 and aplastic sarcoma of the

NOTE Confidence: 0.90234715

01:53:18.195 --> 01:53:20.135 kidney, embryonal rhabdomyosarcomas,

NOTE Confidence: 0.9324544

01:53:20.915 --> 01:53:22.535 and cervical embryonal

NOTE Confidence: 0.94764644

01:53:23.075 --> 01:53:25.500 rhabdomyosarcomas sarcomas are also associated

NOTE Confidence: 0.94764644

01:53:25.500 --> 01:53:26.159 with Dicer

NOTE Confidence: 0.96522355

01:53:26.460 --> 01:53:27.900 one and many other types

NOTE Confidence: 0.96522355

01:53:27.900 --> 01:53:29.360 of tumors including endocrine,

NOTE Confidence: 0.85563815

01:53:29.739 --> 01:53:32.239 CNS tumors, and ovarian tumors,

NOTE Confidence: 0.85563815

01:53:32.260 --> 01:53:33.280 gyn, gynandroblastoma.

NOTE Confidence: 0.98760986

01:53:34.619 --> 01:53:35.900 This is an example of

NOTE Confidence: 0.98760986

01:53:35.900 --> 01:53:36.960 a Dicer one,

NOTE Confidence: 0.99365234

01:53:37.579 --> 01:53:38.079 tumor.
NOTE Confidence: 0.91052246

01:53:38.895 --> 01:53:39.854 You can see is,
NOTE Confidence: 0.96494836

01:53:40.494 --> 01:53:42.255 one of the cystic types
NOTE Confidence: 0.96494836

01:53:42.255 --> 01:53:43.775 not as aggressive but they
NOTE Confidence: 0.96494836

01:53:43.775 --> 01:53:45.474 can be like this
NOTE Confidence: 0.94656575

01:53:46.175 --> 01:53:48.415 or like this or like
NOTE Confidence: 0.94656575

01:53:48.415 --> 01:53:50.755 this. Plural pulmonary blastoma cystic
NOTE Confidence: 0.94656575

01:53:50.814 --> 01:53:52.514 that can be very cystic.
NOTE Confidence: 0.9569782

01:53:53.320 --> 01:53:54.520 This is very difficult to
NOTE Confidence: 0.9569782

01:53:54.520 --> 01:53:56.120 diagnose as a pleural pulmonary
NOTE Confidence: 0.9569782

01:53:56.120 --> 01:53:57.720 blastoma because it looks completely
NOTE Confidence: 0.9569782

01:53:57.720 --> 01:53:59.720 benign and it behaves in
NOTE Confidence: 0.9569782

01:53:59.720 --> 01:54:00.780 a benign fashion.
NOTE Confidence: 0.99408317

01:54:01.240 --> 01:54:02.700 But you have to identify
NOTE Confidence: 0.99408317

01:54:02.760 --> 01:54:04.520 because the patient and the
NOTE Confidence: 0.99408317

01:54:04.520 --> 01:54:06.520 patient's family is at risk

NOTE Confidence: 0.99408317
01:54:06.520 --> 01:54:07.260 of developing
NOTE Confidence: 0.9968262
01:54:07.640 --> 01:54:08.380 other tumors.
NOTE Confidence: 0.9522742
01:54:10.854 --> 01:54:12.135 Here you have a repetition
NOTE Confidence: 0.9522742
01:54:12.135 --> 01:54:13.095 of what I just said.
NOTE Confidence: 0.9522742
01:54:13.095 --> 01:54:14.215 I'm not gonna spend more
NOTE Confidence: 0.9522742
01:54:14.215 --> 01:54:15.575 time here. This is how
NOTE Confidence: 0.9522742
01:54:15.575 --> 01:54:16.695 you pay attention to the
NOTE Confidence: 0.9522742
01:54:16.695 --> 01:54:18.955 stroma and diagnose it as
NOTE Confidence: 0.9522742
01:54:19.015 --> 01:54:21.035 a plural pleural pulmonary blastoma
NOTE Confidence: 0.9522742
01:54:21.095 --> 01:54:22.875 solid with rhabdomyoblastoma
NOTE Confidence: 0.87466866
01:54:23.735 --> 01:54:26.130 like elements with an aplasia,
NOTE Confidence: 0.87466866
01:54:26.189 --> 01:54:27.250 abundant cytoplasm.
NOTE Confidence: 0.96191406
01:54:27.790 --> 01:54:28.830 And then when you proceed
NOTE Confidence: 0.96191406
01:54:28.830 --> 01:54:30.350 with the genetic analysis, you
NOTE Confidence: 0.96191406
01:54:30.350 --> 01:54:32.270 identify the germline Dicer one
NOTE Confidence: 0.96191406

01:54:32.270 --> 01:54:32.770 mutations
NOTE Confidence: 0.89538574
01:54:33.470 --> 01:54:34.770 in, these,
NOTE Confidence: 0.8249512
01:54:35.950 --> 01:54:36.450 tissues.
NOTE Confidence: 0.9209595
01:54:37.065 --> 01:54:38.425 You can also use,
NOTE Confidence: 0.9448242
01:54:38.825 --> 01:54:39.325 immunohistochemistry,
NOTE Confidence: 1
01:54:40.665 --> 01:54:41.165 for
NOTE Confidence: 0.8367513
01:54:42.345 --> 01:54:43.725 labeling these changes.
NOTE Confidence: 0.9317786
01:54:44.105 --> 01:54:44.825 Let me show you a
NOTE Confidence: 0.9317786
01:54:44.825 --> 01:54:46.105 council that I received a
NOTE Confidence: 0.9317786
01:54:46.105 --> 01:54:47.705 few years ago. Came from
NOTE Confidence: 0.9317786
01:54:47.705 --> 01:54:49.385 Italy in a patient of
NOTE Confidence: 0.9317786
01:54:49.385 --> 01:54:50.525 one year old
NOTE Confidence: 0.8815918
01:54:51.349 --> 01:54:51.849 with,
NOTE Confidence: 0.9859314
01:54:52.429 --> 01:54:53.270 I'm sure all of you
NOTE Confidence: 0.9859314
01:54:53.270 --> 01:54:54.250 can read Italian.
NOTE Confidence: 0.88879395
01:54:54.949 --> 01:54:57.449 The patient had medulo epithelioma

NOTE Confidence: 0.993571
01:54:57.829 --> 01:54:58.730 in the eye,
NOTE Confidence: 0.9324544
01:54:59.110 --> 01:54:59.610 retinoblastoma,
NOTE Confidence: 0.94384766
01:55:00.790 --> 01:55:01.290 and,
NOTE Confidence: 0.8630371
01:55:02.230 --> 01:55:03.369 on the one enucleation.
NOTE Confidence: 0.9589178
01:55:03.750 --> 01:55:05.284 So the the patient already
NOTE Confidence: 0.9589178
01:55:05.284 --> 01:55:06.405 had a tumor in the
NOTE Confidence: 0.9589178
01:55:06.405 --> 01:55:06.905 eye,
NOTE Confidence: 0.7972005
01:55:07.284 --> 01:55:07.784 retinoblastoma.
NOTE Confidence: 0.92493784
01:55:08.244 --> 01:55:09.925 It's a relatively common pediatric
NOTE Confidence: 0.92493784
01:55:09.925 --> 01:55:12.165 tumor and then the patient
NOTE Confidence: 0.92493784
01:55:12.165 --> 01:55:12.665 has
NOTE Confidence: 0.9930013
01:55:13.445 --> 01:55:14.505 a long lesion
NOTE Confidence: 0.9407823
01:55:14.885 --> 01:55:16.264 and this is the appearance
NOTE Confidence: 0.9407823
01:55:16.324 --> 01:55:17.685 of the long lesion that
NOTE Confidence: 0.9407823
01:55:17.685 --> 01:55:19.205 I received. These are images
NOTE Confidence: 0.9407823

01:55:19.205 --> 01:55:20.770 taken from that,
NOTE Confidence: 0.8109945

01:55:21.390 --> 01:55:22.610 that's, case.
NOTE Confidence: 0.9414542

01:55:22.990 --> 01:55:24.350 And as you appreciate this
NOTE Confidence: 0.9414542

01:55:24.350 --> 01:55:25.710 cyst and you pay attention
NOTE Confidence: 0.9414542

01:55:25.710 --> 01:55:27.310 to this trauma component of
NOTE Confidence: 0.9414542

01:55:27.310 --> 01:55:28.050 this cyst,
NOTE Confidence: 0.933394

01:55:28.830 --> 01:55:30.910 something is not right and
NOTE Confidence: 0.933394

01:55:30.910 --> 01:55:32.030 then you go in other
NOTE Confidence: 0.933394

01:55:32.030 --> 01:55:32.530 areas,
NOTE Confidence: 0.9510498

01:55:33.230 --> 01:55:35.010 more cellular and then
NOTE Confidence: 0.91430056

01:55:35.585 --> 01:55:37.284 the diagnosis that we issued
NOTE Confidence: 0.91430056

01:55:37.344 --> 01:55:39.045 was this type one cystic
NOTE Confidence: 0.91430056

01:55:39.185 --> 01:55:41.264 pure pulmonary blastoma. We didn't
NOTE Confidence: 0.91430056

01:55:41.264 --> 01:55:42.804 know about the resection margins.
NOTE Confidence: 0.9479806

01:55:43.105 --> 01:55:44.065 We said that there was
NOTE Confidence: 0.9479806

01:55:44.065 --> 01:55:45.185 no involvement of the plural

NOTE Confidence: 0.9479806
01:55:45.185 --> 01:55:46.485 in the material reviewed
NOTE Confidence: 0.86257327
01:55:46.864 --> 01:55:49.045 and the background long parenchyma
NOTE Confidence: 0.94299316
01:55:49.425 --> 01:55:50.485 looked normal.
NOTE Confidence: 0.90249294
01:55:51.240 --> 01:55:52.520 And we made a comment
NOTE Confidence: 0.90249294
01:55:52.520 --> 01:55:54.120 here and referred to the
NOTE Confidence: 0.90249294
01:55:54.120 --> 01:55:56.200 pure, to the beats five
NOTE Confidence: 0.90249294
01:55:56.200 --> 01:55:57.320 edition of the,
NOTE Confidence: 0.9871582
01:55:57.880 --> 01:55:59.580 pediatric tumors in the WHO.
NOTE Confidence: 0.9820731
01:56:01.320 --> 01:56:02.440 Let me show you another
NOTE Confidence: 0.9820731
01:56:02.440 --> 01:56:03.720 example. This is a two
NOTE Confidence: 0.9820731
01:56:03.720 --> 01:56:05.205 year old with a left
NOTE Confidence: 0.9820731
01:56:05.205 --> 01:56:06.965 pneumothorax that you can appreciate
NOTE Confidence: 0.9820731
01:56:06.965 --> 01:56:07.465 here.
NOTE Confidence: 0.99902344
01:56:07.845 --> 01:56:08.505 And then
NOTE Confidence: 0.96275496
01:56:09.285 --> 01:56:10.725 similar lesions to the one
NOTE Confidence: 0.96275496

01:56:10.725 --> 01:56:11.865 that I just discussed,
NOTE Confidence: 0.9773837

01:56:12.725 --> 01:56:15.705 they resected these lesions. Unfortunately,
NOTE Confidence: 0.9773837

01:56:16.005 --> 01:56:18.725 the specimen perforated and it
NOTE Confidence: 0.9773837

01:56:18.725 --> 01:56:19.225 deflated.
NOTE Confidence: 0.9703505

01:56:19.580 --> 01:56:21.120 It was a cystic lesion
NOTE Confidence: 0.9703505

01:56:21.260 --> 01:56:23.660 that appears collapsed here. This
NOTE Confidence: 0.9703505

01:56:23.660 --> 01:56:25.260 is the ink that, our,
NOTE Confidence: 0.9703505

01:56:25.660 --> 01:56:26.960 residents or PAs,
NOTE Confidence: 0.9550781

01:56:27.820 --> 01:56:29.360 dissecting this, example
NOTE Confidence: 0.95513046

01:56:29.740 --> 01:56:31.260 put on to, evaluate the
NOTE Confidence: 0.95513046

01:56:31.260 --> 01:56:32.780 margins. And then when we
NOTE Confidence: 0.95513046

01:56:32.780 --> 01:56:33.760 look at the histology,
NOTE Confidence: 0.9716797

01:56:34.355 --> 01:56:35.875 there are areas like normal
NOTE Confidence: 0.9716797

01:56:35.875 --> 01:56:38.215 lung, areas with little cysts,
NOTE Confidence: 0.9716797

01:56:38.355 --> 01:56:39.554 but then there are areas
NOTE Confidence: 0.9716797

01:56:39.554 --> 01:56:40.295 like this

NOTE Confidence: 0.8721924
01:56:41.715 --> 01:56:42.534 and this.
NOTE Confidence: 0.95422363
01:56:42.835 --> 01:56:44.375 So this is not,
NOTE Confidence: 0.95583636
01:56:46.400 --> 01:56:48.000 as formerly referred to a
NOTE Confidence: 0.95583636
01:56:48.000 --> 01:56:49.520 CPAM. This is a pleural
NOTE Confidence: 0.95583636
01:56:49.520 --> 01:56:51.440 pulmonary blastoma, and you have
NOTE Confidence: 0.95583636
01:56:51.440 --> 01:56:52.719 to pay attention to this
NOTE Confidence: 0.95583636
01:56:52.719 --> 01:56:54.480 because that is what is
NOTE Confidence: 0.95583636
01:56:54.480 --> 01:56:56.400 going to determine the progression
NOTE Confidence: 0.95583636
01:56:56.400 --> 01:56:57.520 of the disease and the
NOTE Confidence: 0.95583636
01:56:57.520 --> 01:56:59.679 treatment necessary to control this
NOTE Confidence: 0.95583636
01:56:59.679 --> 01:57:01.514 entity. There is an aplasia
NOTE Confidence: 0.95583636
01:57:01.574 --> 01:57:03.815 in this stromal cells, very
NOTE Confidence: 0.95583636
01:57:03.815 --> 01:57:05.414 significant an aplasia in some
NOTE Confidence: 0.95583636
01:57:05.414 --> 01:57:06.934 areas, and we issued a
NOTE Confidence: 0.95583636
01:57:06.934 --> 01:57:08.614 diagnosis. Actually, this was a
NOTE Confidence: 0.95583636

01:57:08.614 --> 01:57:09.815 case that there was not
NOTE Confidence: 0.95583636

01:57:09.815 --> 01:57:10.855 mine, but it was in
NOTE Confidence: 0.95583636

01:57:10.855 --> 01:57:12.155 my department. And the,
NOTE Confidence: 0.94895583

01:57:12.695 --> 01:57:14.474 the all these cases were
NOTE Confidence: 0.94895583

01:57:14.534 --> 01:57:15.750 shared with us, and they
NOTE Confidence: 0.94895583

01:57:15.750 --> 01:57:16.730 they used to
NOTE Confidence: 0.85530597

01:57:17.190 --> 01:57:18.390 incorporate our opinions on the
NOTE Confidence: 0.85530597

01:57:18.390 --> 01:57:20.470 pathology signing this case, call
NOTE Confidence: 0.85530597

01:57:20.470 --> 01:57:22.470 it pluripotent blastoma type two
NOTE Confidence: 0.85530597

01:57:22.470 --> 01:57:24.150 because it's not the very
NOTE Confidence: 0.85530597

01:57:24.150 --> 01:57:24.650 cystic
NOTE Confidence: 0.9032157

01:57:24.950 --> 01:57:26.230 but not the very solid.
NOTE Confidence: 0.9032157

01:57:26.230 --> 01:57:27.050 It's mixed
NOTE Confidence: 0.73271835

01:57:27.350 --> 01:57:29.670 and then multiple multifocal an
NOTE Confidence: 0.73271835

01:57:29.670 --> 01:57:30.955 aplasia, a drabdomyomatous
NOTE Confidence: 0.8633034

01:57:32.135 --> 01:57:34.155 differentiation with multifocal an aplasia,

NOTE Confidence: 0.8633034
01:57:34.375 --> 01:57:36.135 etcetera, etcetera, and they comment
NOTE Confidence: 0.8633034
01:57:36.135 --> 01:57:36.635 here.
NOTE Confidence: 0.99031574
01:57:40.855 --> 01:57:41.835 How about this?
NOTE Confidence: 0.9894206
01:57:42.455 --> 01:57:43.255 We are not in the
NOTE Confidence: 0.9894206
01:57:43.255 --> 01:57:43.755 lung.
NOTE Confidence: 0.98922527
01:57:44.450 --> 01:57:46.310 This is a cystic lesion.
NOTE Confidence: 0.96850586
01:57:47.330 --> 01:57:48.950 You recognize this. Right?
NOTE Confidence: 0.890625
01:57:49.890 --> 01:57:50.630 The testis
NOTE Confidence: 0.90145874
01:57:51.250 --> 01:57:52.310 is in the paratesticular
NOTE Confidence: 1
01:57:53.410 --> 01:57:53.910 area.
NOTE Confidence: 0.99934894
01:57:54.610 --> 01:57:55.670 I put my
NOTE Confidence: 0.9406128
01:57:56.050 --> 01:57:58.070 cell phone under the glass
NOTE Confidence: 0.9406128
01:57:58.370 --> 01:57:59.350 taking the picture
NOTE Confidence: 0.9225586
01:57:59.725 --> 01:58:01.905 to see the trans illumination
NOTE Confidence: 0.9976129
01:58:02.285 --> 01:58:03.185 of the cyst.
NOTE Confidence: 0.9626465

01:58:03.645 --> 01:58:04.145 Okay?
NOTE Confidence: 0.9888071

01:58:04.685 --> 01:58:06.125 And, yeah, residents laugh at
NOTE Confidence: 0.9888071

01:58:06.125 --> 01:58:07.085 me when I do these
NOTE Confidence: 0.9888071

01:58:07.085 --> 01:58:07.985 kinds of things
NOTE Confidence: 0.9753418

01:58:08.364 --> 01:58:09.485 because they say, why did
NOTE Confidence: 0.9753418

01:58:09.485 --> 01:58:10.844 you do that? Well, I
NOTE Confidence: 0.9753418

01:58:10.844 --> 01:58:11.725 want to see the trans
NOTE Confidence: 0.9753418

01:58:11.725 --> 01:58:13.005 illumination. Right? And and you
NOTE Confidence: 0.9753418

01:58:13.005 --> 01:58:13.985 just have to
NOTE Confidence: 0.9846802

01:58:14.760 --> 01:58:16.300 follow your imagination. And,
NOTE Confidence: 0.9958984

01:58:17.320 --> 01:58:19.020 when when I did this,
NOTE Confidence: 0.91097003

01:58:19.719 --> 01:58:21.579 the fellow that was rotating
NOTE Confidence: 0.91097003

01:58:21.719 --> 01:58:23.079 and doing his fellowship with
NOTE Confidence: 0.91097003

01:58:23.079 --> 01:58:24.599 us in Pittsburgh, doctor Louis
NOTE Confidence: 0.91097003

01:58:24.599 --> 01:58:25.980 Samson, a fantastic,
NOTE Confidence: 0.92244464

01:58:26.520 --> 01:58:28.695 fellow, pediatric pathology fellow from

NOTE Confidence: 0.92244464

01:58:28.695 --> 01:58:30.295 Canada, he put this,

NOTE Confidence: 0.9219749

01:58:32.055 --> 01:58:33.335 pictures in the tumor board,

NOTE Confidence: 0.9219749

01:58:33.335 --> 01:58:34.375 and this was the final

NOTE Confidence: 0.9219749

01:58:34.375 --> 01:58:34.875 diagnosis.

NOTE Confidence: 0.9256657

01:58:35.335 --> 01:58:38.215 Cystically dilated epididymal ducts consisting

NOTE Confidence: 0.9256657

01:58:38.215 --> 01:58:39.895 with obstructive changes. Yes. Yes.

NOTE Confidence: 0.9256657

01:58:39.895 --> 01:58:41.415 Yes. And then low grade

NOTE Confidence: 0.9256657

01:58:41.415 --> 01:58:43.035 myxoid spindle cell neoplasm

NOTE Confidence: 0.9201172

01:58:43.495 --> 01:58:45.060 with a comment. And we

NOTE Confidence: 0.9201172

01:58:45.060 --> 01:58:46.580 learned that in the clinical

NOTE Confidence: 0.9201172

01:58:46.580 --> 01:58:48.500 history there was a history

NOTE Confidence: 0.9201172

01:58:48.500 --> 01:58:51.000 of Dicer one syndrome. So

NOTE Confidence: 0.9201172

01:58:51.060 --> 01:58:53.320 this lesion fits perfectly well.

NOTE Confidence: 0.9807193

01:58:53.860 --> 01:58:54.980 The fact that we saw

NOTE Confidence: 0.9807193

01:58:54.980 --> 01:58:56.420 it after the patient has

NOTE Confidence: 0.9807193

01:58:56.420 --> 01:58:58.280 been diagnosed with Dicer one
NOTE Confidence: 0.9807193

01:58:58.555 --> 01:58:59.995 is good, but it doesn't
NOTE Confidence: 0.9807193

01:58:59.995 --> 01:59:01.835 matter. It fits everything. And
NOTE Confidence: 0.9807193

01:59:01.835 --> 01:59:03.195 if you see this before
NOTE Confidence: 0.9807193

01:59:03.195 --> 01:59:05.115 knowing the clinical history, you
NOTE Confidence: 0.9807193

01:59:05.115 --> 01:59:06.335 are after it.
NOTE Confidence: 0.83979493

01:59:07.835 --> 01:59:10.175 Recently, Estero Olivas group in,
NOTE Confidence: 0.96899414

01:59:10.640 --> 01:59:11.680 that evil place of the
NOTE Confidence: 0.96899414

01:59:11.680 --> 01:59:12.180 north,
NOTE Confidence: 0.8881836

01:59:12.640 --> 01:59:14.340 published these series of cases
NOTE Confidence: 0.85025024

01:59:14.800 --> 01:59:16.580 with Dicer one related
NOTE Confidence: 0.6432292

01:59:16.960 --> 01:59:17.460 WIMS
NOTE Confidence: 0.9116211

01:59:17.920 --> 01:59:18.500 like uterine
NOTE Confidence: 0.798584

01:59:18.800 --> 01:59:19.300 tumor,
NOTE Confidence: 0.9885905

01:59:20.160 --> 01:59:22.160 with, several cases reported. And
NOTE Confidence: 0.9885905

01:59:22.160 --> 01:59:24.020 the the series are growing

NOTE Confidence: 0.9885905
01:59:24.225 --> 01:59:26.165 in adults and in pediatric
NOTE Confidence: 0.82349855
01:59:26.465 --> 01:59:28.725 examples of dysent one mutations.
NOTE Confidence: 0.99869794
01:59:31.585 --> 01:59:33.445 Let me switch to another
NOTE Confidence: 0.99869794
01:59:33.585 --> 01:59:34.085 interesting
NOTE Confidence: 1
01:59:34.465 --> 01:59:34.965 finding.
NOTE Confidence: 0.97329533
01:59:35.345 --> 01:59:36.465 This is a five year
NOTE Confidence: 0.97329533
01:59:36.465 --> 01:59:38.130 old girl with a palpable
NOTE Confidence: 0.97329533
01:59:38.130 --> 01:59:39.489 suprapubic mass. This is a
NOTE Confidence: 0.97329533
01:59:39.489 --> 01:59:40.310 Yale case
NOTE Confidence: 0.96583337
01:59:41.250 --> 01:59:42.929 that, came to us with
NOTE Confidence: 0.96583337
01:59:42.929 --> 01:59:45.330 two independent masses. And, when
NOTE Confidence: 0.96583337
01:59:45.330 --> 01:59:46.370 I was looking at this
NOTE Confidence: 0.96583337
01:59:46.370 --> 01:59:47.830 case, I said two masses.
NOTE Confidence: 0.96583337
01:59:47.969 --> 01:59:49.590 And I remember doctor McNamara,
NOTE Confidence: 0.9743768
01:59:50.225 --> 01:59:51.665 Joe McNamara, one of the
NOTE Confidence: 0.9743768

01:59:51.665 --> 01:59:52.645 pediatric oncologists,
NOTE Confidence: 0.9128529

01:59:53.505 --> 01:59:55.425 told me, yeah. Our surgeons
NOTE Confidence: 0.9128529

01:59:55.425 --> 01:59:56.625 think that this is a
NOTE Confidence: 0.9128529

01:59:56.625 --> 01:59:58.225 one's tumor that dropped down
NOTE Confidence: 0.9128529

01:59:58.225 --> 01:59:59.905 and metastasis into the bladder.
NOTE Confidence: 0.9128529

01:59:59.905 --> 02:00:01.365 I had never seen that
NOTE Confidence: 0.9128529

02:00:01.425 --> 02:00:02.545 and I have not seen
NOTE Confidence: 0.9128529

02:00:02.545 --> 02:00:04.005 that still now.
NOTE Confidence: 0.94331056

02:00:04.920 --> 02:00:05.960 So five year old with
NOTE Confidence: 0.94331056

02:00:05.960 --> 02:00:08.380 two tumors, there was, however,
NOTE Confidence: 0.98079425

02:00:09.400 --> 02:00:11.260 developmental delay in the clinical
NOTE Confidence: 0.98079425

02:00:11.400 --> 02:00:11.900 history.
NOTE Confidence: 0.9975586

02:00:12.360 --> 02:00:13.820 And because of that
NOTE Confidence: 0.76626587

02:00:14.280 --> 02:00:16.150 and facial deep dysmorphism,
NOTE Confidence: 0.98136395

02:00:17.365 --> 02:00:19.065 a karyotype was obtained
NOTE Confidence: 0.9671056

02:00:19.445 --> 02:00:20.885 at that time. And they

NOTE Confidence: 0.9671056
02:00:20.885 --> 02:00:22.805 found an interstitial deletion of
NOTE Confidence: 0.9671056
02:00:22.805 --> 02:00:24.485 nine q twenty two q
NOTE Confidence: 0.9671056
02:00:24.485 --> 02:00:25.385 thirty two.
NOTE Confidence: 0.99188703
02:00:26.485 --> 02:00:27.525 And that part of the
NOTE Confidence: 0.99188703
02:00:27.525 --> 02:00:29.465 clinical history made me think
NOTE Confidence: 0.99188703
02:00:29.525 --> 02:00:30.585 in something else.
NOTE Confidence: 0.9721755
02:00:32.260 --> 02:00:33.620 In addition, there was mild
NOTE Confidence: 0.9721755
02:00:33.620 --> 02:00:34.840 hypertrophy, hemihypertrophy,
NOTE Confidence: 0.99853516
02:00:35.380 --> 02:00:36.360 prominent forehead,
NOTE Confidence: 0.7624756
02:00:36.980 --> 02:00:37.480 hypertilarism,
NOTE Confidence: 0.97264713
02:00:37.860 --> 02:00:39.860 epicanthal folds, etcetera, etcetera, etcetera,
NOTE Confidence: 0.97264713
02:00:39.860 --> 02:00:41.320 all that you read there.
NOTE Confidence: 0.97264713
02:00:41.460 --> 02:00:42.820 These are the images of
NOTE Confidence: 0.97264713
02:00:42.820 --> 02:00:44.040 the kidney tumor
NOTE Confidence: 1
02:00:44.805 --> 02:00:45.305 with
NOTE Confidence: 0.78479004

02:00:45.844 --> 02:00:46.665 a epithelial
NOTE Confidence: 0.6953125

02:00:47.925 --> 02:00:48.425 slash,
NOTE Confidence: 0.5813802

02:00:48.885 --> 02:00:49.385 blastema,
NOTE Confidence: 0.90528286

02:00:50.245 --> 02:00:52.165 components in the Wilms tumor
NOTE Confidence: 0.90528286

02:00:52.165 --> 02:00:53.285 that looks a little weird,
NOTE Confidence: 0.90528286

02:00:53.285 --> 02:00:54.265 but it is
NOTE Confidence: 0.9230957

02:00:54.645 --> 02:00:55.785 a Wilms tumor.
NOTE Confidence: 0.9942491

02:00:56.645 --> 02:00:58.085 And these are the images
NOTE Confidence: 0.9942491

02:00:58.085 --> 02:00:59.305 of the bladder too.
NOTE Confidence: 0.9502991

02:01:00.000 --> 02:01:01.380 If you see them separately,
NOTE Confidence: 0.9502991

02:01:01.679 --> 02:01:03.300 you will diagnose one's tumor
NOTE Confidence: 0.9486287

02:01:03.679 --> 02:01:05.360 next, and you will diagnose
NOTE Confidence: 0.9486287

02:01:05.360 --> 02:01:05.860 rhabdomyosarcoma
NOTE Confidence: 0.9711914

02:01:06.960 --> 02:01:07.460 next.
NOTE Confidence: 0.9324254

02:01:08.159 --> 02:01:10.239 Here, we have stroma with
NOTE Confidence: 0.9324254

02:01:10.239 --> 02:01:10.900 an aplasia,

NOTE Confidence: 0.74580896
02:01:11.679 --> 02:01:12.900 desmin, myogenic
NOTE Confidence: 0.8964844
02:01:13.599 --> 02:01:14.099 expression,
NOTE Confidence: 0.9043045
02:01:14.515 --> 02:01:15.335 classic rhabdomyosarcoma.
NOTE Confidence: 0.9475911
02:01:16.355 --> 02:01:17.475 But the patient has two
NOTE Confidence: 0.9475911
02:01:17.475 --> 02:01:18.595 tumors and is five years
NOTE Confidence: 0.9475911
02:01:18.595 --> 02:01:19.255 of age.
NOTE Confidence: 0.9941406
02:01:19.795 --> 02:01:20.835 If you see that in
NOTE Confidence: 0.9941406
02:01:20.835 --> 02:01:21.495 a sixty
NOTE Confidence: 0.987264
02:01:21.955 --> 02:01:24.035 something years old patient, two
NOTE Confidence: 0.987264
02:01:24.035 --> 02:01:24.535 tumors
NOTE Confidence: 0.9036458
02:01:24.995 --> 02:01:26.295 is very common
NOTE Confidence: 0.7858073
02:01:26.675 --> 02:01:27.495 in a kid.
NOTE Confidence: 0.9926758
02:01:27.955 --> 02:01:28.455 No.
NOTE Confidence: 0.98423076
02:01:30.240 --> 02:01:32.080 So when I rereview the
NOTE Confidence: 0.98423076
02:01:32.080 --> 02:01:33.840 clinical history and I saw
NOTE Confidence: 0.98423076

02:01:33.840 --> 02:01:35.940 the interstitial deletion in chromosome
NOTE Confidence: 0.98423076

02:01:36.000 --> 02:01:38.160 nine, I went to my
NOTE Confidence: 0.98423076

02:01:38.160 --> 02:01:38.980 late wife's
NOTE Confidence: 0.9502441

02:01:39.280 --> 02:01:41.380 lab in the genetics department
NOTE Confidence: 0.9502441

02:01:41.600 --> 02:01:43.540 working with doctor Alan Bale.
NOTE Confidence: 0.9807617

02:01:44.375 --> 02:01:46.155 Myra was my late wife,
NOTE Confidence: 0.96623677

02:01:46.695 --> 02:01:47.975 and she was working in
NOTE Confidence: 0.96623677

02:01:47.975 --> 02:01:49.975 that laboratory. And Alan Bale
NOTE Confidence: 0.96623677

02:01:49.975 --> 02:01:51.094 happens to be the person
NOTE Confidence: 0.96623677

02:01:51.094 --> 02:01:51.915 that identify
NOTE Confidence: 0.99902344

02:01:52.455 --> 02:01:53.275 the patched
NOTE Confidence: 0.85180664

02:01:53.735 --> 02:01:54.235 gene.
NOTE Confidence: 0.99471027

02:01:55.094 --> 02:01:57.150 And the patched gene is
NOTE Confidence: 0.99471027

02:01:57.150 --> 02:01:57.650 responsible
NOTE Confidence: 1

02:01:58.110 --> 02:01:58.610 for
NOTE Confidence: 0.27661133

02:01:58.910 --> 02:01:59.410 Goling

NOTE Confidence: 0.9020996
02:01:59.870 --> 02:02:00.370 syndrome,
NOTE Confidence: 0.9977539
02:02:00.910 --> 02:02:02.449 which is a cancer predisposition
NOTE Confidence: 0.84277344
02:02:02.910 --> 02:02:03.410 syndrome,
NOTE Confidence: 0.9962158
02:02:03.790 --> 02:02:05.650 a very important cancer predisposition
NOTE Confidence: 0.9962158
02:02:05.870 --> 02:02:07.410 syndrome. You can
NOTE Confidence: 0.9274224
02:02:08.445 --> 02:02:10.045 advise the clinicians to pursue
NOTE Confidence: 0.9274224
02:02:10.045 --> 02:02:11.645 the diagnosis of Golgi syndrome
NOTE Confidence: 0.9274224
02:02:11.645 --> 02:02:13.025 when you receive an odontogenic
NOTE Confidence: 0.9507324
02:02:13.325 --> 02:02:15.665 keratocyst because they are commonly
NOTE Confidence: 0.99869794
02:02:16.205 --> 02:02:17.885 the first red flag to
NOTE Confidence: 0.99869794
02:02:17.885 --> 02:02:18.385 identify
NOTE Confidence: 0.97505695
02:02:18.925 --> 02:02:20.445 a Golgi syndrome in a
NOTE Confidence: 0.97505695
02:02:20.445 --> 02:02:20.945 child.
NOTE Confidence: 0.5185547
02:02:22.350 --> 02:02:22.850 Syndrome
NOTE Confidence: 0.88239396
02:02:23.230 --> 02:02:25.570 described by, doctor Robert Golin
NOTE Confidence: 0.88239396

02:02:25.630 --> 02:02:26.450 in Minneapolis
NOTE Confidence: 0.81778973

02:02:27.390 --> 02:02:29.090 is, a multi,
NOTE Confidence: 0.66292316

02:02:30.750 --> 02:02:31.250 tumoral,
NOTE Confidence: 0.9558919

02:02:32.030 --> 02:02:33.730 cancer predisposition syndrome,
NOTE Confidence: 0.97648114

02:02:34.670 --> 02:02:36.210 with, three genes
NOTE Confidence: 0.92610586

02:02:36.675 --> 02:02:38.295 right now that are associated
NOTE Confidence: 0.92610586

02:02:38.355 --> 02:02:40.435 with, with it. Patch one
NOTE Confidence: 0.92610586

02:02:40.435 --> 02:02:42.515 is the first one. Sufu
NOTE Confidence: 0.92610586

02:02:42.515 --> 02:02:43.555 is another one and then
NOTE Confidence: 0.92610586

02:02:43.555 --> 02:02:44.595 patch two. This is the
NOTE Confidence: 0.92610586

02:02:44.595 --> 02:02:45.715 one that Alan Bale, and
NOTE Confidence: 0.92610586

02:02:45.715 --> 02:02:47.395 this is seventeen to ninety
NOTE Confidence: 0.92610586

02:02:47.395 --> 02:02:48.915 percent of cases are associated
NOTE Confidence: 0.92610586

02:02:48.915 --> 02:02:50.135 with patch one abnormalities.
NOTE Confidence: 0.78999835

02:02:51.340 --> 02:02:53.020 We published this space this
NOTE Confidence: 0.78999835

02:02:53.020 --> 02:02:53.520 case,

NOTE Confidence: 0.78356063

02:02:54.140 --> 02:02:56.560 as a combination of randomized

NOTE Confidence: 0.78356063

02:02:56.620 --> 02:02:58.540 sarcoma, windstorm, and deletion of

NOTE Confidence: 0.78356063

02:02:58.540 --> 02:03:00.160 the patching in syndrome

NOTE Confidence: 0.9256134

02:03:00.699 --> 02:03:02.080 appears in nature oncology,

NOTE Confidence: 0.93670654

02:03:02.620 --> 02:03:03.739 and it has been,

NOTE Confidence: 0.7380371

02:03:04.940 --> 02:03:05.440 important,

NOTE Confidence: 0.94990236

02:03:06.105 --> 02:03:07.705 interesting paper in my in

NOTE Confidence: 0.94990236

02:03:07.705 --> 02:03:10.125 in my, personal collection because,

NOTE Confidence: 0.9982422

02:03:10.745 --> 02:03:12.285 it taught me a lot.

NOTE Confidence: 0.986921

02:03:12.585 --> 02:03:13.865 You can see multiple basal

NOTE Confidence: 0.986921

02:03:13.865 --> 02:03:14.685 cell carcinomas

NOTE Confidence: 0.87895507

02:03:15.065 --> 02:03:16.265 in these tumors in these

NOTE Confidence: 0.87895507

02:03:16.265 --> 02:03:17.625 patients, and this is how

NOTE Confidence: 0.87895507

02:03:17.625 --> 02:03:19.645 Robert Gordon, Bob Gordon identified

NOTE Confidence: 0.90405273

02:03:20.105 --> 02:03:21.870 the the syndrome. He was

NOTE Confidence: 0.9926758

02:03:22.250 --> 02:03:23.150 on a bus
NOTE Confidence: 0.95318604

02:03:24.090 --> 02:03:26.010 riding in Minneapolis and saw
NOTE Confidence: 0.95318604

02:03:26.010 --> 02:03:27.650 a patient, and he used
NOTE Confidence: 0.95318604

02:03:27.650 --> 02:03:29.530 to go and say, excuse
NOTE Confidence: 0.95318604

02:03:29.530 --> 02:03:30.910 me. I am doctor Gordon.
NOTE Confidence: 0.9656513

02:03:31.290 --> 02:03:32.170 Do you mind if I
NOTE Confidence: 0.9656513

02:03:32.170 --> 02:03:33.050 speak with you for a
NOTE Confidence: 0.9656513

02:03:33.050 --> 02:03:33.950 for a minute?
NOTE Confidence: 0.825236

02:03:34.635 --> 02:03:36.075 They will be, no. No.
NOTE Confidence: 0.825236

02:03:36.075 --> 02:03:36.575 Sure.
NOTE Confidence: 0.973565

02:03:36.955 --> 02:03:37.995 And then he will explain
NOTE Confidence: 0.973565

02:03:37.995 --> 02:03:39.275 that he's concerned with these
NOTE Confidence: 0.973565

02:03:39.275 --> 02:03:40.635 lesions that the patient has
NOTE Confidence: 0.973565

02:03:40.635 --> 02:03:41.535 in his face.
NOTE Confidence: 0.9163662

02:03:42.155 --> 02:03:44.255 And then he described the
NOTE Confidence: 0.9163662

02:03:44.395 --> 02:03:45.755 the the the deletion. This

NOTE Confidence: 0.9163662

02:03:45.755 --> 02:03:46.655 is Bob Golin.

NOTE Confidence: 0.88545734

02:03:47.470 --> 02:03:49.310 We invited Bob Golin to

NOTE Confidence: 0.88545734

02:03:49.310 --> 02:03:50.670 come to give Grand Rounds

NOTE Confidence: 0.88545734

02:03:50.670 --> 02:03:52.270 here many years ago, a

NOTE Confidence: 0.88545734

02:03:52.270 --> 02:03:53.710 couple years up, before he

NOTE Confidence: 0.88545734

02:03:53.710 --> 02:03:54.210 said.

NOTE Confidence: 0.99902344

02:03:54.990 --> 02:03:56.770 And when I invited him,

NOTE Confidence: 0.93874514

02:03:57.710 --> 02:03:59.490 I called his office directly,

NOTE Confidence: 0.9851888

02:04:00.285 --> 02:04:01.585 and, the secretary

NOTE Confidence: 0.9231703

02:04:02.205 --> 02:04:04.225 answered and said and I

NOTE Confidence: 0.9231703

02:04:04.445 --> 02:04:05.565 said, this is doctor Reyes

NOTE Confidence: 0.9231703

02:04:05.565 --> 02:04:06.765 from Yale University. May I

NOTE Confidence: 0.9231703

02:04:06.765 --> 02:04:07.965 speak with doctor Gorin? I

NOTE Confidence: 0.9231703

02:04:07.965 --> 02:04:09.645 am I am narrating this

NOTE Confidence: 0.9231703

02:04:09.645 --> 02:04:11.645 conversation here. And and she

NOTE Confidence: 0.9231703

02:04:11.645 --> 02:04:12.980 goes, yes. Just a minute,
NOTE Confidence: 0.9231703
02:04:12.980 --> 02:04:13.480 please.
NOTE Confidence: 0.95476073
02:04:13.860 --> 02:04:14.740 And then she comes to
NOTE Confidence: 0.95476073
02:04:14.740 --> 02:04:15.960 the phone and I said,
NOTE Confidence: 0.9663086
02:04:16.260 --> 02:04:16.900 she says,
NOTE Confidence: 0.9346381
02:04:17.380 --> 02:04:18.820 hello? And I said, doctor
NOTE Confidence: 0.9346381
02:04:18.820 --> 02:04:19.320 Gorlin,
NOTE Confidence: 0.7392578
02:04:19.780 --> 02:04:20.280 si
NOTE Confidence: 0.8432617
02:04:20.660 --> 02:04:21.400 in Spanish.
NOTE Confidence: 0.970166
02:04:22.020 --> 02:04:23.640 Okay? I switched to Spanish
NOTE Confidence: 0.92488605
02:04:24.020 --> 02:04:25.540 and I start speaking with
NOTE Confidence: 0.92488605
02:04:25.540 --> 02:04:26.040 him
NOTE Confidence: 0.8000488
02:04:26.420 --> 02:04:27.160 and then
NOTE Confidence: 0.94962907
02:04:27.695 --> 02:04:29.135 I switched back to English,
NOTE Confidence: 0.94962907
02:04:29.135 --> 02:04:30.255 and I I invited him
NOTE Confidence: 0.94962907
02:04:30.255 --> 02:04:30.895 and all that. And I

NOTE Confidence: 0.94962907
02:04:30.895 --> 02:04:32.255 said, doctor Corin, where did
NOTE Confidence: 0.94962907
02:04:32.255 --> 02:04:34.035 you learn your Spanish?
NOTE Confidence: 0.9282715
02:04:36.415 --> 02:04:37.375 He told me that he
NOTE Confidence: 0.9282715
02:04:37.375 --> 02:04:39.235 was part of a CIA
NOTE Confidence: 0.7114258
02:04:40.175 --> 02:04:40.755 CIA mission
NOTE Confidence: 0.9686279
02:04:42.320 --> 02:04:43.780 in the Nordic countries
NOTE Confidence: 0.8308919
02:04:44.880 --> 02:04:46.800 and that he spent six
NOTE Confidence: 0.8308919
02:04:46.800 --> 02:04:47.300 weeks
NOTE Confidence: 0.99902344
02:04:47.840 --> 02:04:48.340 sharing
NOTE Confidence: 0.96376956
02:04:48.800 --> 02:04:51.140 the room with a Spanish
NOTE Confidence: 0.99902344
02:04:51.760 --> 02:04:52.260 priest
NOTE Confidence: 0.9956665
02:04:52.800 --> 02:04:53.840 that was part of the
NOTE Confidence: 0.9956665
02:04:53.840 --> 02:04:55.220 same CIA mission.
NOTE Confidence: 0.98573524
02:04:55.905 --> 02:04:56.785 I didn't want to know
NOTE Confidence: 0.98573524
02:04:56.785 --> 02:04:57.685 anything else.
NOTE Confidence: 0.9685407

02:04:59.265 --> 02:05:00.545 I didn't bear to ask
NOTE Confidence: 0.9685407

02:05:00.545 --> 02:05:01.445 anything else.
NOTE Confidence: 0.9251874

02:05:02.225 --> 02:05:03.525 She she was a fantastic
NOTE Confidence: 0.9251874

02:05:03.585 --> 02:05:06.065 individual, enormous guy. And when
NOTE Confidence: 0.9251874

02:05:06.065 --> 02:05:07.265 he came into my office,
NOTE Confidence: 0.9251874

02:05:07.265 --> 02:05:09.580 he barely fit. And, he's,
NOTE Confidence: 0.9251874

02:05:09.900 --> 02:05:10.780 told me that he had
NOTE Confidence: 0.9251874

02:05:10.780 --> 02:05:12.960 mantle cell lymphoma. He said,
NOTE Confidence: 0.9251874

02:05:13.020 --> 02:05:14.540 my spleen is the size
NOTE Confidence: 0.9251874

02:05:14.540 --> 02:05:15.280 of Texas.
NOTE Confidence: 0.9812622

02:05:15.820 --> 02:05:17.100 He died a few years
NOTE Confidence: 0.9812622

02:05:17.100 --> 02:05:18.140 later, but he was a
NOTE Confidence: 0.9812622

02:05:18.140 --> 02:05:19.200 fantastic individual.
NOTE Confidence: 0.93640137

02:05:21.100 --> 02:05:22.080 Another example,
NOTE Confidence: 0.9516696

02:05:22.845 --> 02:05:24.364 bloom syndrome. And this is
NOTE Confidence: 0.9516696

02:05:24.364 --> 02:05:26.445 something that I also saw

NOTE Confidence: 0.9516696
02:05:26.445 --> 02:05:27.345 here at Yale.
NOTE Confidence: 0.94020236
02:05:28.205 --> 02:05:30.045 Dan, Pat, Jane was my
NOTE Confidence: 0.94020236
02:05:30.045 --> 02:05:31.805 resident, was a resident at
NOTE Confidence: 0.94020236
02:05:31.805 --> 02:05:32.845 that time. I'm sure that
NOTE Confidence: 0.94020236
02:05:32.845 --> 02:05:34.605 you know who doctor Jane,
NOTE Confidence: 0.94020236
02:05:34.605 --> 02:05:36.785 famous doctor Dampa Jane is.
NOTE Confidence: 0.94020236
02:05:37.085 --> 02:05:38.465 And we saw this
NOTE Confidence: 0.92719185
02:05:41.060 --> 02:05:42.980 patient with a bloom syndrome
NOTE Confidence: 0.92719185
02:05:42.980 --> 02:05:44.360 and a Williams tumor.
NOTE Confidence: 0.96606445
02:05:44.740 --> 02:05:46.340 This is a syndrome that,
NOTE Confidence: 0.96606445
02:05:46.900 --> 02:05:47.400 involves
NOTE Confidence: 0.78201294
02:05:47.860 --> 02:05:49.960 repairing DNA, repairing enzymes.
NOTE Confidence: 0.98312986
02:05:50.500 --> 02:05:51.800 And when they are broken,
NOTE Confidence: 0.98312986
02:05:52.020 --> 02:05:53.265 they don't repair the DNA.
NOTE Confidence: 0.96637374
02:05:53.825 --> 02:05:55.685 This case had an aplasia
NOTE Confidence: 0.8477783

02:05:56.305 --> 02:05:57.525 as you can see here
NOTE Confidence: 0.8477783

02:05:57.745 --> 02:05:58.245 and
NOTE Confidence: 0.9951172

02:05:58.865 --> 02:05:59.685 the sister
NOTE Confidence: 0.98323566

02:06:00.145 --> 02:06:01.125 of this patient
NOTE Confidence: 0.93778485

02:06:01.425 --> 02:06:02.645 developed a hepatocellular
NOTE Confidence: 0.9511719

02:06:03.105 --> 02:06:03.605 carcinoma.
NOTE Confidence: 0.97249347

02:06:04.625 --> 02:06:06.805 Both had the same genetic
NOTE Confidence: 0.97249347

02:06:06.945 --> 02:06:07.445 abnormalities.
NOTE Confidence: 0.99731445

02:06:08.290 --> 02:06:09.570 Two children in the same
NOTE Confidence: 0.99731445

02:06:09.570 --> 02:06:11.030 family with two
NOTE Confidence: 0.99902344

02:06:11.409 --> 02:06:11.909 relatively
NOTE Confidence: 0.90893555

02:06:12.210 --> 02:06:13.350 infrequent lesions.
NOTE Confidence: 0.99934894

02:06:13.889 --> 02:06:14.949 They need to
NOTE Confidence: 0.953125

02:06:15.489 --> 02:06:16.929 rise the red flag for
NOTE Confidence: 0.953125

02:06:16.929 --> 02:06:18.610 you to pursue it. The
NOTE Confidence: 0.953125

02:06:18.610 --> 02:06:20.370 pathologist cannot be just sitting

NOTE Confidence: 0.953125
02:06:20.370 --> 02:06:21.670 in the office and making,
NOTE Confidence: 0.9637553
02:06:22.155 --> 02:06:23.835 as my wife says, when
NOTE Confidence: 0.9637553
02:06:23.835 --> 02:06:25.195 when we are working together,
NOTE Confidence: 0.9637553
02:06:25.195 --> 02:06:26.395 she says, oh, you are
NOTE Confidence: 0.9637553
02:06:26.395 --> 02:06:27.675 just giving names to these
NOTE Confidence: 0.9637553
02:06:27.675 --> 02:06:29.035 animals, right, to these,
NOTE Confidence: 0.9815918
02:06:29.915 --> 02:06:31.915 cows. Well, no. We we
NOTE Confidence: 0.9815918
02:06:31.915 --> 02:06:33.135 are part of the clinical
NOTE Confidence: 0.9921875
02:06:34.500 --> 02:06:35.000 advisory
NOTE Confidence: 0.9735541
02:06:35.860 --> 02:06:37.720 team for the patient's family.
NOTE Confidence: 0.9735541
02:06:38.020 --> 02:06:39.620 We need to pursue that
NOTE Confidence: 0.9735541
02:06:39.620 --> 02:06:40.980 they follow it because if
NOTE Confidence: 0.9735541
02:06:40.980 --> 02:06:42.020 they if you don't do
NOTE Confidence: 0.9735541
02:06:42.020 --> 02:06:43.300 that, they may or may
NOTE Confidence: 0.9735541
02:06:43.300 --> 02:06:44.900 not realize that it is
NOTE Confidence: 0.9735541

02:06:44.900 --> 02:06:45.400 important.
NOTE Confidence: 0.9958496

02:06:47.065 --> 02:06:47.864 I don't know if I
NOTE Confidence: 0.9958496

02:06:47.864 --> 02:06:48.184 would,
NOTE Confidence: 0.9946777

02:06:48.665 --> 02:06:50.364 repeat the diagnosis of hepatocellular
NOTE Confidence: 0.95494443

02:06:50.665 --> 02:06:52.264 carcinoma on this particular case
NOTE Confidence: 0.95494443

02:06:52.264 --> 02:06:54.425 because hepatocellular carcinoma in children
NOTE Confidence: 0.95494443

02:06:54.425 --> 02:06:56.025 is relatively rare and is
NOTE Confidence: 0.95494443

02:06:56.025 --> 02:06:57.565 in a process of being
NOTE Confidence: 0.95494443

02:06:57.864 --> 02:06:58.364 reclassified
NOTE Confidence: 0.92317325

02:06:58.985 --> 02:07:00.610 as we speak in the
NOTE Confidence: 0.92317325

02:07:00.610 --> 02:07:02.130 sixth edition of the of
NOTE Confidence: 0.92317325

02:07:02.130 --> 02:07:04.630 the WHO Digestive Tumors book.
NOTE Confidence: 0.92317325

02:07:04.770 --> 02:07:06.290 But we published these these
NOTE Confidence: 0.92317325

02:07:06.290 --> 02:07:06.790 patients
NOTE Confidence: 0.79467773

02:07:07.330 --> 02:07:08.550 with bloom syndrome
NOTE Confidence: 0.7141113

02:07:08.930 --> 02:07:09.250 with,

NOTE Confidence: 0.8539551
02:07:09.810 --> 02:07:10.310 heparocellular
NOTE Confidence: 0.9459519
02:07:10.770 --> 02:07:12.290 and WIMS tumor in both
NOTE Confidence: 0.9459519
02:07:12.290 --> 02:07:13.170 of them in the same
NOTE Confidence: 0.9459519
02:07:13.170 --> 02:07:15.215 family. So those are clues
NOTE Confidence: 0.9459519
02:07:15.275 --> 02:07:17.034 you see the same family
NOTE Confidence: 0.9459519
02:07:17.034 --> 02:07:18.815 having two children with tumors.
NOTE Confidence: 0.9459519
02:07:19.114 --> 02:07:20.175 That's not frequent.
NOTE Confidence: 0.91758895
02:07:22.155 --> 02:07:24.235 Bloom syndrome looks like this
NOTE Confidence: 0.91758895
02:07:24.235 --> 02:07:26.175 clinically. They have hypersensitivity
NOTE Confidence: 0.94262695
02:07:26.795 --> 02:07:27.300 to the,
NOTE Confidence: 0.8527832
02:07:28.020 --> 02:07:28.520 sunlight,
NOTE Confidence: 0.97542316
02:07:28.900 --> 02:07:30.520 and they have multiple abnormal
NOTE Confidence: 0.97542316
02:07:30.820 --> 02:07:32.280 abnormalities in the karyotypes
NOTE Confidence: 0.96657985
02:07:32.659 --> 02:07:34.440 due to the abnormal mutations
NOTE Confidence: 0.96657985
02:07:34.579 --> 02:07:36.579 in the repair enzymes of
NOTE Confidence: 0.96657985

02:07:36.579 --> 02:07:38.360 DNA. I'm not gonna detail
NOTE Confidence: 0.96657985

02:07:38.420 --> 02:07:39.480 all these because
NOTE Confidence: 0.8607178

02:07:39.860 --> 02:07:40.980 as we are gonna run
NOTE Confidence: 0.8607178

02:07:40.980 --> 02:07:41.800 out of time.
NOTE Confidence: 0.98733604

02:07:43.014 --> 02:07:45.175 I'm gonna probably finish with,
NOTE Confidence: 0.98733604

02:07:45.494 --> 02:07:47.335 this group of disorders, multiple
NOTE Confidence: 0.98733604

02:07:47.335 --> 02:07:48.954 endocrine neoplasia syndromes.
NOTE Confidence: 0.97147214

02:07:49.414 --> 02:07:50.695 These are typically seen in
NOTE Confidence: 0.97147214

02:07:50.695 --> 02:07:52.295 adults. We all are familiar
NOTE Confidence: 0.97147214

02:07:52.295 --> 02:07:53.414 with them but you can
NOTE Confidence: 0.97147214

02:07:53.414 --> 02:07:54.795 also see them in children.
NOTE Confidence: 0.8598633

02:07:56.780 --> 02:07:58.320 This individual here
NOTE Confidence: 0.8654785

02:07:58.940 --> 02:08:00.800 is doctor Aiden Carney.
NOTE Confidence: 0.9559733

02:08:01.660 --> 02:08:02.940 I'm sure that many of
NOTE Confidence: 0.9559733

02:08:02.940 --> 02:08:04.700 you have heard of the
NOTE Confidence: 0.9559733

02:08:04.700 --> 02:08:05.760 Kearney syndrome,

NOTE Confidence: 0.95703125

02:08:06.460 --> 02:08:07.520 Kearney's triad,

NOTE Confidence: 0.9242452

02:08:08.014 --> 02:08:10.275 etcetera, etcetera. Kearney's complex.

NOTE Confidence: 0.9561523

02:08:10.655 --> 02:08:12.114 He described all of those.

NOTE Confidence: 0.9705241

02:08:12.815 --> 02:08:14.014 He is sitting with me

NOTE Confidence: 0.9705241

02:08:14.014 --> 02:08:15.215 in my office here at

NOTE Confidence: 0.9705241

02:08:15.215 --> 02:08:17.074 Yale about twenty years ago.

NOTE Confidence: 0.9549235

02:08:17.454 --> 02:08:19.235 I invited him twice. Once

NOTE Confidence: 0.9549235

02:08:19.295 --> 02:08:20.895 here and once in Pittsburgh.

NOTE Confidence: 0.9549235

02:08:20.895 --> 02:08:22.355 He was a fantastic guy.

NOTE Confidence: 0.86938477

02:08:22.735 --> 02:08:23.235 Small,

NOTE Confidence: 0.9232178

02:08:24.090 --> 02:08:25.389 Irish gentleman,

NOTE Confidence: 0.9501953

02:08:25.929 --> 02:08:27.389 funny, and very,

NOTE Confidence: 0.99858946

02:08:27.850 --> 02:08:29.150 you know, combative.

NOTE Confidence: 0.99853516

02:08:29.770 --> 02:08:30.429 I remember

NOTE Confidence: 0.8751221

02:08:31.130 --> 02:08:32.489 I I remember that,

NOTE Confidence: 0.8718262

02:08:33.210 --> 02:08:34.489 I think Stuart Flynn,
NOTE Confidence: 0.93436104

02:08:34.810 --> 02:08:35.850 told me that he was
NOTE Confidence: 0.93436104

02:08:35.850 --> 02:08:37.530 presenting something, and then Alan
NOTE Confidence: 0.93436104

02:08:37.530 --> 02:08:39.175 Carney was jumping in the
NOTE Confidence: 0.93436104

02:08:39.175 --> 02:08:40.855 audience trying to contradict him
NOTE Confidence: 0.93436104

02:08:40.855 --> 02:08:42.375 and he found him, a
NOTE Confidence: 0.93436104

02:08:42.375 --> 02:08:43.755 very combative spirit.
NOTE Confidence: 0.942749

02:08:44.695 --> 02:08:46.295 Very nice guy. And,
NOTE Confidence: 0.8537089

02:08:47.415 --> 02:08:48.614 we are referring to the
NOTE Confidence: 0.8537089

02:08:48.614 --> 02:08:50.074 Sippel Williams Pierce,
NOTE Confidence: 0.9526367

02:08:50.390 --> 02:08:51.270 triad of,
NOTE Confidence: 0.9069498

02:08:51.750 --> 02:08:53.530 individuals that describe the multiple
NOTE Confidence: 0.9069498

02:08:53.590 --> 02:08:56.730 endocrine neoplasia syndromes where had
NOTE Confidence: 0.9980469

02:08:57.190 --> 02:08:58.250 a major role.
NOTE Confidence: 0.97965086

02:08:58.630 --> 02:08:59.750 There are many types of
NOTE Confidence: 0.97965086

02:08:59.750 --> 02:09:02.250 these syndromes, cancer predisposition syndromes.

NOTE Confidence: 0.9637451

02:09:02.790 --> 02:09:04.230 And what happened with,

NOTE Confidence: 0.986084

02:09:05.675 --> 02:09:07.055 in in in this particular,

NOTE Confidence: 0.9954427

02:09:07.435 --> 02:09:08.795 group of disorders is that

NOTE Confidence: 0.9954427

02:09:08.795 --> 02:09:10.495 we saw two sisters

NOTE Confidence: 0.9628906

02:09:11.275 --> 02:09:12.335 many years ago

NOTE Confidence: 0.9941406

02:09:12.795 --> 02:09:13.695 that came

NOTE Confidence: 0.9995117

02:09:14.475 --> 02:09:15.375 at five

NOTE Confidence: 0.93362087

02:09:15.995 --> 02:09:18.075 years, eleven months, and three

NOTE Confidence: 0.93362087

02:09:18.075 --> 02:09:19.775 years and eight months,

NOTE Confidence: 0.9953613

02:09:20.520 --> 02:09:22.619 Both to be reviewed

NOTE Confidence: 0.9996745

02:09:22.920 --> 02:09:23.820 by the clinicians

NOTE Confidence: 0.98898655

02:09:24.280 --> 02:09:25.400 that were looking at the

NOTE Confidence: 0.98898655

02:09:25.400 --> 02:09:27.260 thyroid because they had

NOTE Confidence: 0.99609375

02:09:27.639 --> 02:09:28.139 a

NOTE Confidence: 0.9887044

02:09:28.520 --> 02:09:30.619 family history of thyroid carcinoma.

NOTE Confidence: 0.9327741

02:09:31.400 --> 02:09:33.000 And I happen to receive

NOTE Confidence: 0.9327741

02:09:33.000 --> 02:09:33.659 the biopsies.

NOTE Confidence: 0.9190956

02:09:34.215 --> 02:09:35.735 We put them together. We

NOTE Confidence: 0.9190956

02:09:35.735 --> 02:09:36.855 we still publish in black

NOTE Confidence: 0.9190956

02:09:36.855 --> 02:09:37.755 and white in

NOTE Confidence: 0.8565267

02:09:38.135 --> 02:09:40.135 those those years and this

NOTE Confidence: 0.8565267

02:09:40.135 --> 02:09:40.635 individual

NOTE Confidence: 0.94628906

02:09:41.895 --> 02:09:43.815 is relatively well known, Jim

NOTE Confidence: 0.94628906

02:09:43.815 --> 02:09:44.315 Gill,

NOTE Confidence: 0.98950195

02:09:44.855 --> 02:09:46.295 whose wife is sitting right

NOTE Confidence: 0.98950195

02:09:46.295 --> 02:09:46.795 here.

NOTE Confidence: 0.9712469

02:09:47.095 --> 02:09:49.539 She was my resident also,

NOTE Confidence: 0.9712469

02:09:49.679 --> 02:09:51.039 and we put together this

NOTE Confidence: 0.9712469

02:09:51.039 --> 02:09:51.539 paper

NOTE Confidence: 0.76627606

02:09:52.079 --> 02:09:53.059 and we advise

NOTE Confidence: 0.99902344
02:09:53.599 --> 02:09:54.340 to perform
NOTE Confidence: 0.998291
02:09:54.880 --> 02:09:55.380 prophylactic
NOTE Confidence: 0.9499512
02:09:55.920 --> 02:09:56.420 thyroidectomy
NOTE Confidence: 0.99902344
02:09:57.360 --> 02:09:58.099 in young
NOTE Confidence: 0.92659134
02:09:58.400 --> 02:10:00.239 children. And, look, this this
NOTE Confidence: 0.92659134
02:10:00.239 --> 02:10:01.280 is an advice that is
NOTE Confidence: 0.92659134
02:10:01.280 --> 02:10:01.780 very
NOTE Confidence: 0.93393177
02:10:02.715 --> 02:10:04.075 delicate to me because you
NOTE Confidence: 0.93393177
02:10:04.075 --> 02:10:05.115 go to the parents and
NOTE Confidence: 0.93393177
02:10:05.115 --> 02:10:06.575 you say, look,
NOTE Confidence: 0.9033203
02:10:07.595 --> 02:10:08.575 you have to
NOTE Confidence: 0.8876321
02:10:08.955 --> 02:10:10.315 do daughters. One of them
NOTE Confidence: 0.8876321
02:10:10.315 --> 02:10:12.575 has unfortunately medullary carcinoma,
NOTE Confidence: 0.993103
02:10:13.035 --> 02:10:14.315 but the other one has
NOTE Confidence: 0.993103
02:10:14.315 --> 02:10:15.535 this change that
NOTE Confidence: 0.9540364

02:10:16.780 --> 02:10:17.900 food seems is going to
NOTE Confidence: 0.9540364

02:10:17.900 --> 02:10:19.900 progress to medullary carcinoma. So
NOTE Confidence: 0.9540364

02:10:19.900 --> 02:10:21.260 you need to recognize that
NOTE Confidence: 0.9540364

02:10:21.260 --> 02:10:22.780 and be part of that
NOTE Confidence: 0.9540364

02:10:22.780 --> 02:10:24.300 exercise in discussing with the
NOTE Confidence: 0.9540364

02:10:24.300 --> 02:10:25.900 patients. We frequently speak with
NOTE Confidence: 0.9540364

02:10:25.900 --> 02:10:27.280 patients. Claudia is a pediatrician,
NOTE Confidence: 0.9540364

02:10:27.340 --> 02:10:28.380 so she she doesn't have
NOTE Confidence: 0.9540364

02:10:28.380 --> 02:10:29.260 a problem with that, but
NOTE Confidence: 0.9540364

02:10:29.260 --> 02:10:30.320 I am not a clinician.
NOTE Confidence: 0.9778878

02:10:30.705 --> 02:10:31.744 But I love to speak
NOTE Confidence: 0.9778878

02:10:31.744 --> 02:10:33.105 with the patients and advise
NOTE Confidence: 0.9778878

02:10:33.105 --> 02:10:34.625 them in conjunction with our
NOTE Confidence: 0.9778878

02:10:34.625 --> 02:10:37.344 clinical colleagues in pediatric oncology.
NOTE Confidence: 0.9778878

02:10:37.344 --> 02:10:39.105 So we published this tale
NOTE Confidence: 0.9778878

02:10:39.105 --> 02:10:39.925 of two sisters

NOTE Confidence: 0.9836426
02:10:40.625 --> 02:10:41.025 with,
NOTE Confidence: 0.9797363
02:10:41.505 --> 02:10:42.324 Jim Gill
NOTE Confidence: 0.9031937
02:10:42.640 --> 02:10:44.160 and doctor Miron Janelle, who
NOTE Confidence: 0.9031937
02:10:44.160 --> 02:10:45.540 was a pediatric endocrinologist
NOTE Confidence: 0.9239705
02:10:46.160 --> 02:10:47.840 at that time directing the
NOTE Confidence: 0.9239705
02:10:47.840 --> 02:10:49.840 program of thyroid cancer in
NOTE Confidence: 0.9239705
02:10:49.840 --> 02:10:50.739 Yale University,
NOTE Confidence: 0.99746096
02:10:51.120 --> 02:10:52.420 and it was a fantastic
NOTE Confidence: 0.9904785
02:10:52.720 --> 02:10:53.220 experience.
NOTE Confidence: 0.88710535
02:10:54.985 --> 02:10:56.825 Multiple endocrine neoplasia has many
NOTE Confidence: 0.88710535
02:10:56.825 --> 02:10:58.525 different expressions, histologically
NOTE Confidence: 0.95288086
02:10:58.985 --> 02:11:01.405 speaking. It has multiple neurofibromas,
NOTE Confidence: 0.9366862
02:11:01.945 --> 02:11:03.945 neuromas, etcetera and you have
NOTE Confidence: 0.9366862
02:11:03.945 --> 02:11:04.525 to be
NOTE Confidence: 0.99348956
02:11:04.905 --> 02:11:05.885 able to recognize
NOTE Confidence: 0.9738206

02:11:06.265 --> 02:11:08.105 the danger that exists when
NOTE Confidence: 0.9738206

02:11:08.105 --> 02:11:09.625 you see lesions like this
NOTE Confidence: 0.9738206

02:11:09.625 --> 02:11:10.680 in young children.
NOTE Confidence: 0.97845817

02:11:11.000 --> 02:11:12.680 Young children should not get
NOTE Confidence: 0.97845817

02:11:12.680 --> 02:11:14.460 sick and if they do,
NOTE Confidence: 0.97845817

02:11:14.520 --> 02:11:16.440 they usually get well with
NOTE Confidence: 0.97845817

02:11:16.440 --> 02:11:17.180 no help
NOTE Confidence: 0.9421313

02:11:17.720 --> 02:11:19.240 but when they don't, you
NOTE Confidence: 0.9421313

02:11:19.240 --> 02:11:20.920 need to pay attention to
NOTE Confidence: 0.9421313

02:11:20.920 --> 02:11:21.420 them.
NOTE Confidence: 0.9373101

02:11:22.360 --> 02:11:23.640 I had a few more
NOTE Confidence: 0.9373101

02:11:23.640 --> 02:11:24.920 examples that I am not
NOTE Confidence: 0.9373101

02:11:24.920 --> 02:11:26.284 going to detail because I'm
NOTE Confidence: 0.9373101

02:11:26.284 --> 02:11:27.165 not going to,
NOTE Confidence: 0.9130046

02:11:27.725 --> 02:11:28.945 spend, well, maybe,
NOTE Confidence: 0.9984375

02:11:29.405 --> 02:11:30.364 just to show you that

NOTE Confidence: 0.9984375

02:11:30.364 --> 02:11:32.065 there are several other associations

NOTE Confidence: 0.9166667

02:11:32.844 --> 02:11:34.525 of involving the genital syndrome.

NOTE Confidence: 0.9166667

02:11:34.525 --> 02:11:36.545 For example, the genital area,

NOTE Confidence: 0.9166667

02:11:36.605 --> 02:11:38.685 for example, Fraser syndrome where

NOTE Confidence: 0.9166667

02:11:38.685 --> 02:11:40.045 you can have mutations in

NOTE Confidence: 0.9166667

02:11:40.045 --> 02:11:40.545 WT

NOTE Confidence: 0.5690918

02:11:41.090 --> 02:11:41.490 one,

NOTE Confidence: 0.9522569

02:11:41.890 --> 02:11:43.730 gene with different types of

NOTE Confidence: 0.9522569

02:11:43.730 --> 02:11:45.570 mutation that can lead to

NOTE Confidence: 0.9522569

02:11:45.570 --> 02:11:46.710 different syndromes

NOTE Confidence: 0.9431234

02:11:47.170 --> 02:11:48.610 depending on the codon that

NOTE Confidence: 0.9431234

02:11:48.610 --> 02:11:50.130 is involved and you can

NOTE Confidence: 0.9431234

02:11:50.130 --> 02:11:52.150 have abnormalities in the kidney

NOTE Confidence: 0.949349

02:11:52.530 --> 02:11:54.050 and in the gonads. You

NOTE Confidence: 0.949349

02:11:54.050 --> 02:11:54.950 can have gonadoblastoma

NOTE Confidence: 0.9306216

02:11:55.865 --> 02:11:57.545 like this. When you see
NOTE Confidence: 0.9306216

02:11:57.545 --> 02:11:59.305 different the differences in sex
NOTE Confidence: 0.9306216

02:11:59.305 --> 02:12:00.905 development like a strict gonad
NOTE Confidence: 0.9306216

02:12:00.905 --> 02:12:02.585 that you see here associated
NOTE Confidence: 0.9306216

02:12:02.585 --> 02:12:03.405 with a gonadoblastoma
NOTE Confidence: 0.9463806

02:12:04.585 --> 02:12:06.345 and then you pursue the
NOTE Confidence: 0.9463806

02:12:06.345 --> 02:12:07.705 genetics, you can,
NOTE Confidence: 0.9911133

02:12:08.425 --> 02:12:10.685 diagnose a cancer predisposition syndrome.
NOTE Confidence: 1

02:12:12.239 --> 02:12:12.739 So
NOTE Confidence: 0.9492853

02:12:13.280 --> 02:12:15.280 the major non CNS pediatric
NOTE Confidence: 0.9492853

02:12:15.280 --> 02:12:17.940 tumor categories are, mentioned here.
NOTE Confidence: 0.9492853

02:12:18.079 --> 02:12:19.280 Most of them I have
NOTE Confidence: 0.9492853

02:12:19.280 --> 02:12:20.719 touched on but I want
NOTE Confidence: 0.9492853

02:12:20.719 --> 02:12:21.199 you to,
NOTE Confidence: 0.9904785

02:12:22.159 --> 02:12:23.619 take home these messages.
NOTE Confidence: 0.99455565

02:12:24.905 --> 02:12:26.665 Ten percent of pediatric solid

NOTE Confidence: 0.99455565
02:12:26.665 --> 02:12:29.245 tumors represent cancer predisposition syndromes.
NOTE Confidence: 0.99967444
02:12:29.705 --> 02:12:30.445 The histopathology
NOTE Confidence: 0.9918213
02:12:31.145 --> 02:12:33.085 frequently gives you the sentinel
NOTE Confidence: 0.9918213
02:12:33.145 --> 02:12:33.645 clue
NOTE Confidence: 0.9835612
02:12:33.945 --> 02:12:35.385 to start working on those
NOTE Confidence: 0.9835612
02:12:35.385 --> 02:12:35.885 patients.
NOTE Confidence: 0.99520874
02:12:36.745 --> 02:12:38.185 You have to correlate genetics
NOTE Confidence: 0.99520874
02:12:38.185 --> 02:12:39.405 with clinical data.
NOTE Confidence: 0.87736005
02:12:40.239 --> 02:12:41.139 You, the pathologist
NOTE Confidence: 0.98183596
02:12:41.599 --> 02:12:43.940 identifies patients at risk frequently
NOTE Confidence: 0.91646904
02:12:44.800 --> 02:12:46.000 as long as you think
NOTE Confidence: 0.91646904
02:12:46.000 --> 02:12:46.659 of this.
NOTE Confidence: 0.9730573
02:12:47.040 --> 02:12:48.420 You can just pass it
NOTE Confidence: 0.9730573
02:12:48.560 --> 02:12:50.340 one more slide and next
NOTE Confidence: 0.9730573
02:12:50.480 --> 02:12:51.699 and you miss it.
NOTE Confidence: 0.99365234

02:12:52.095 --> 02:12:53.235 And collaboration
NOTE Confidence: 0.9324544

02:12:53.614 --> 02:12:54.515 with our clinical,
NOTE Confidence: 0.989624

02:12:54.975 --> 02:12:56.195 genetic, oncological,
NOTE Confidence: 0.9411621

02:12:56.895 --> 02:12:57.395 surgical,
NOTE Confidence: 0.81396484

02:12:58.255 --> 02:12:59.475 pediatric specialists
NOTE Confidence: 0.95703125

02:12:59.935 --> 02:13:00.435 assures
NOTE Confidence: 0.9951172

02:13:00.815 --> 02:13:01.315 adequate
NOTE Confidence: 0.98291016

02:13:01.935 --> 02:13:03.235 precision diagnosis.
NOTE Confidence: 0.9379127

02:13:03.775 --> 02:13:05.040 Okay? So there is a
NOTE Confidence: 0.9379127

02:13:05.040 --> 02:13:06.639 great variety great variety of
NOTE Confidence: 0.9379127

02:13:06.639 --> 02:13:09.040 syndromes predisposing to cancer that
NOTE Confidence: 0.9379127

02:13:09.040 --> 02:13:10.980 appear in childhood, the pediatric
NOTE Confidence: 0.9379127

02:13:11.040 --> 02:13:11.540 pathologist,
NOTE Confidence: 0.95064604

02:13:12.000 --> 02:13:13.460 the first to detect suspicious
NOTE Confidence: 0.95064604

02:13:13.520 --> 02:13:15.040 or diagnostic findings in these
NOTE Confidence: 0.95064604

02:13:15.040 --> 02:13:16.420 syndromes, not infrequently,

NOTE Confidence: 0.9467882

02:13:16.915 --> 02:13:18.995 And the developing organism has

NOTE Confidence: 0.9467882

02:13:18.995 --> 02:13:20.855 highly variable ways of anotypic

NOTE Confidence: 0.9467882

02:13:20.995 --> 02:13:23.335 expression with the pediatric pathologist,

NOTE Confidence: 0.79125977

02:13:24.114 --> 02:13:24.935 which with

NOTE Confidence: 0.867296

02:13:25.395 --> 02:13:27.014 the with which the pediatric

NOTE Confidence: 0.867296

02:13:27.074 --> 02:13:28.775 pathologist should be intimately

NOTE Confidence: 0.97204775

02:13:29.170 --> 02:13:31.269 familiarized. If you don't recognize

NOTE Confidence: 0.97204775

02:13:31.329 --> 02:13:32.849 these lesions, you miss an

NOTE Confidence: 0.97204775

02:13:32.849 --> 02:13:34.710 important diagnosis that will affect

NOTE Confidence: 0.97204775

02:13:34.769 --> 02:13:36.130 not only that patient, but

NOTE Confidence: 0.97204775

02:13:36.130 --> 02:13:37.110 the entire family.

NOTE Confidence: 0.9489746

02:13:38.449 --> 02:13:40.070 And I finish with that.

NOTE Confidence: 0.93387043

02:13:42.545 --> 02:13:44.385 You may recognize yourself, some

NOTE Confidence: 0.93387043

02:13:44.385 --> 02:13:46.385 of yourselves here. I couldn't

NOTE Confidence: 0.93387043

02:13:46.385 --> 02:13:48.324 put everyone but, you know,

NOTE Confidence: 0.99609375

02:13:49.025 --> 02:13:49.525 nice
NOTE Confidence: 0.79508466

02:13:50.145 --> 02:13:51.905 memories from Jim Gill, Marie
NOTE Confidence: 0.79508466

02:13:51.905 --> 02:13:52.405 Robert,
NOTE Confidence: 0.8757324

02:13:52.864 --> 02:13:53.185 and,
NOTE Confidence: 0.9930943

02:13:54.170 --> 02:13:55.130 well, I am not going
NOTE Confidence: 0.9930943

02:13:55.130 --> 02:13:56.810 to name everyone because you
NOTE Confidence: 0.9930943

02:13:56.810 --> 02:13:58.030 know who they are.
NOTE Confidence: 0.98706055

02:13:58.330 --> 02:13:59.530 Thank you very much for
NOTE Confidence: 0.98706055

02:13:59.530 --> 02:14:01.210 the honor of, visiting Yale
NOTE Confidence: 0.98706055

02:14:01.210 --> 02:14:01.710 again.
NOTE Confidence: 0.9987793

02:14:02.409 --> 02:14:03.150 Thank you.
NOTE Confidence: 0.97631836

02:14:07.385 --> 02:14:08.285 Any questions,
NOTE Confidence: 0.99902344

02:14:08.825 --> 02:14:09.325 comments?
NOTE Confidence: 0.9522027

02:14:13.785 --> 02:14:14.985 I I know that you
NOTE Confidence: 0.9522027

02:14:14.985 --> 02:14:16.205 always have a question.
NOTE Confidence: 0.7326276

02:14:19.850 --> 02:14:20.958 You know what they do?

NOTE Confidence: 0.7326276
02:14:20.958 --> 02:14:22.406 You need sequencing or sequence,
NOTE Confidence: 0.7326276
02:14:22.406 --> 02:14:23.853 whatever, like, all it is
NOTE Confidence: 0.7326276
02:14:23.853 --> 02:14:25.301 is that they need to
NOTE Confidence: 0.7326276
02:14:25.301 --> 02:14:26.570 walk in sequence. Can you
NOTE Confidence: 0.7326276
02:14:26.570 --> 02:14:27.370 share with us what do
NOTE Confidence: 0.7326276
02:14:27.370 --> 02:14:28.330 you think are driving the
NOTE Confidence: 0.7326276
02:14:28.330 --> 02:14:29.850 next just a lot of
NOTE Confidence: 0.7326276
02:14:29.850 --> 02:14:31.390 things we don't have recognized
NOTE Confidence: 0.7326276
02:14:31.530 --> 02:14:32.570 that are gonna come out
NOTE Confidence: 0.7326276
02:14:32.570 --> 02:14:33.470 of all that sequence?
NOTE Confidence: 0.99902344
02:14:34.534 --> 02:14:35.034 Yeah.
NOTE Confidence: 0.9964844
02:14:35.335 --> 02:14:36.635 It's a it's a very
NOTE Confidence: 0.9599753
02:14:37.175 --> 02:14:38.855 important point, John, that you
NOTE Confidence: 0.9599753
02:14:38.855 --> 02:14:40.375 raised. And, believe me, it's
NOTE Confidence: 0.9599753
02:14:40.375 --> 02:14:41.895 a very hot point of
NOTE Confidence: 0.9599753

02:14:41.895 --> 02:14:42.955 discussion now.
NOTE Confidence: 0.9709251

02:14:43.574 --> 02:14:45.094 And Jose and I exchanged
NOTE Confidence: 0.9709251

02:14:45.094 --> 02:14:46.315 a couple of years ago
NOTE Confidence: 0.9709251

02:14:46.375 --> 02:14:48.429 papers on the relevance of
NOTE Confidence: 0.9709251

02:14:48.429 --> 02:14:49.949 phenotype and genetic changes and
NOTE Confidence: 0.9709251

02:14:49.949 --> 02:14:50.530 the correlation
NOTE Confidence: 0.97473145

02:14:50.989 --> 02:14:52.670 of these two things. And
NOTE Confidence: 0.97473145

02:14:52.670 --> 02:14:54.349 right now, the WHO is
NOTE Confidence: 0.97473145

02:14:54.349 --> 02:14:55.409 suffering from,
NOTE Confidence: 0.96365213

02:14:57.150 --> 02:14:58.510 I wouldn't say attacks, but
NOTE Confidence: 0.96365213

02:14:58.510 --> 02:15:00.349 questioning, severe questioning from our
NOTE Confidence: 0.96365213

02:15:00.349 --> 02:15:02.270 clinical colleagues that say, well,
NOTE Confidence: 0.96365213

02:15:02.270 --> 02:15:04.165 you know, histology doesn't matter
NOTE Confidence: 0.96365213

02:15:04.165 --> 02:15:04.665 anymore.
NOTE Confidence: 0.9739091

02:15:05.045 --> 02:15:06.505 You just need to sequence.
NOTE Confidence: 0.9739091

02:15:06.645 --> 02:15:07.525 You just need to put

NOTE Confidence: 0.9739091
02:15:07.525 --> 02:15:10.005 the patient's tissue through, genetic
NOTE Confidence: 0.9739091
02:15:10.005 --> 02:15:11.865 analysis and identify deletion.
NOTE Confidence: 0.96897197
02:15:12.565 --> 02:15:13.925 And I wish that could
NOTE Confidence: 0.96897197
02:15:13.925 --> 02:15:15.545 be possible but it isn't
NOTE Confidence: 0.96897197
02:15:15.605 --> 02:15:16.105 because
NOTE Confidence: 0.9970703
02:15:18.739 --> 02:15:21.080 many different types of genetic
NOTE Confidence: 0.9970703
02:15:21.140 --> 02:15:21.640 abnormalities
NOTE Confidence: 0.9977214
02:15:22.020 --> 02:15:23.160 that are identifiable
NOTE Confidence: 0.976888
02:15:23.699 --> 02:15:26.180 by sequencing different modalities of
NOTE Confidence: 0.976888
02:15:26.180 --> 02:15:26.680 sequencing
NOTE Confidence: 0.81591797
02:15:27.620 --> 02:15:28.120 generate
NOTE Confidence: 0.9996745
02:15:28.660 --> 02:15:30.120 completely different phenotypes
NOTE Confidence: 0.9737142
02:15:30.820 --> 02:15:32.100 depending on a number of
NOTE Confidence: 0.9737142
02:15:32.100 --> 02:15:32.600 other
NOTE Confidence: 0.9794922
02:15:33.864 --> 02:15:34.685 things, epigenetics
NOTE Confidence: 0.8359375

02:15:35.145 --> 02:15:35.965 and environmental
NOTE Confidence: 0.9916992

02:15:36.665 --> 02:15:37.165 circumstances.
NOTE Confidence: 0.8753052

02:15:38.105 --> 02:15:39.864 The phenotype, which is what
NOTE Confidence: 0.8753052

02:15:39.864 --> 02:15:41.324 we pathologists see
NOTE Confidence: 0.77372235

02:15:41.784 --> 02:15:43.085 macro or microscopically,
NOTE Confidence: 0.7394206

02:15:44.585 --> 02:15:45.725 is really the
NOTE Confidence: 0.9458008

02:15:46.610 --> 02:15:47.110 unquestionable
NOTE Confidence: 0.99726564

02:15:47.570 --> 02:15:49.510 end result of the genetic
NOTE Confidence: 1

02:15:50.130 --> 02:15:50.630 expression
NOTE Confidence: 0.9970703

02:15:51.490 --> 02:15:52.710 of an organism.
NOTE Confidence: 0.99726564

02:15:53.330 --> 02:15:54.870 The phenotype of a tissue
NOTE Confidence: 0.9066162

02:15:55.970 --> 02:15:57.830 contains the information to
NOTE Confidence: 0.9995117

02:15:58.215 --> 02:15:59.915 understand how the genetics
NOTE Confidence: 0.98853827

02:16:00.295 --> 02:16:02.055 led to that change, but
NOTE Confidence: 0.98853827

02:16:02.055 --> 02:16:03.435 the reverse is not true.
NOTE Confidence: 0.98853827

02:16:03.735 --> 02:16:05.255 If you only see the

NOTE Confidence: 0.98853827
02:16:05.255 --> 02:16:06.315 part of the genetics,
NOTE Confidence: 0.99912107
02:16:06.935 --> 02:16:08.715 you will miss certain conditions.
NOTE Confidence: 0.99902344
02:16:09.255 --> 02:16:09.755 So
NOTE Confidence: 0.9998779
02:16:10.420 --> 02:16:11.539 I hope that we are
NOTE Confidence: 0.9998779
02:16:11.539 --> 02:16:12.440 able to develop
NOTE Confidence: 0.9903231
02:16:12.820 --> 02:16:14.980 additional instruments on both sides
NOTE Confidence: 0.9903231
02:16:14.980 --> 02:16:16.440 of the operation, the genetic
NOTE Confidence: 0.9903231
02:16:16.659 --> 02:16:17.159 analysis
NOTE Confidence: 0.88183594
02:16:17.619 --> 02:16:18.599 and the histological
NOTE Confidence: 0.8098795
02:16:19.940 --> 02:16:22.679 structural, ultra structural, etcetera analysis
NOTE Confidence: 0.8954102
02:16:23.300 --> 02:16:24.760 to bring this to a
NOTE Confidence: 0.9885254
02:16:25.204 --> 02:16:27.224 uniform reality that can serve
NOTE Confidence: 0.9885254
02:16:27.445 --> 02:16:29.224 and guide us for diagnosis
NOTE Confidence: 0.9885254
02:16:29.284 --> 02:16:31.364 and treatment. This issue was
NOTE Confidence: 0.9885254
02:16:31.364 --> 02:16:32.425 pointed at
NOTE Confidence: 0.93459064

02:16:33.045 --> 02:16:34.325 in a recent paper that
NOTE Confidence: 0.93459064

02:16:34.325 --> 02:16:35.685 we, the editorial board of
NOTE Confidence: 0.93459064

02:16:35.685 --> 02:16:36.345 the WHO
NOTE Confidence: 0.89746094

02:16:37.040 --> 02:16:37.540 books
NOTE Confidence: 0.87482375

02:16:37.920 --> 02:16:39.700 published in the Lancet Oncology
NOTE Confidence: 0.87482375

02:16:40.000 --> 02:16:41.760 Journal a few months ago,
NOTE Confidence: 0.87482375

02:16:41.760 --> 02:16:42.500 last year,
NOTE Confidence: 0.9480366

02:16:42.879 --> 02:16:44.080 where we made the case
NOTE Confidence: 0.9480366

02:16:44.080 --> 02:16:45.840 that, yes, we understand that
NOTE Confidence: 0.9480366

02:16:45.840 --> 02:16:47.600 genetic analysis is more and
NOTE Confidence: 0.9480366

02:16:47.600 --> 02:16:48.820 more and more important,
NOTE Confidence: 0.9973958

02:16:49.200 --> 02:16:50.260 but we cannot
NOTE Confidence: 0.9980469

02:16:50.640 --> 02:16:51.140 eliminate
NOTE Confidence: 0.9566586

02:16:51.985 --> 02:16:53.345 the the role of the
NOTE Confidence: 0.9566586

02:16:53.345 --> 02:16:55.504 histological analysis because we miss
NOTE Confidence: 0.9566586

02:16:55.504 --> 02:16:57.745 the phenotype changes and we

NOTE Confidence: 0.9566586
02:16:57.745 --> 02:16:59.845 miss the precision diagnosis.
NOTE Confidence: 0.9849772
02:17:00.945 --> 02:17:01.745 So I don't know if
NOTE Confidence: 0.9849772
02:17:01.745 --> 02:17:03.504 that is a very wishy
NOTE Confidence: 0.9849772
02:17:03.504 --> 02:17:05.525 washy response to you, but
NOTE Confidence: 0.95703125
02:17:07.660 --> 02:17:08.160 We
NOTE Confidence: 0.9838867
02:17:08.900 --> 02:17:09.400 we
NOTE Confidence: 0.7703334
02:17:10.580 --> 02:17:12.360 whole career has been identifying
NOTE Confidence: 0.7703334
02:17:12.580 --> 02:17:13.320 new lesions.
NOTE Confidence: 0.9337891
02:17:14.180 --> 02:17:15.300 Now you have a huge
NOTE Confidence: 0.9337891
02:17:15.300 --> 02:17:17.160 resource in the genomic database,
NOTE Confidence: 0.97324914
02:17:18.100 --> 02:17:19.620 and so it should be
NOTE Confidence: 0.97324914
02:17:19.620 --> 02:17:20.280 a pointer.
NOTE Confidence: 0.77793974
02:17:21.145 --> 02:17:21.865 But we have to look
NOTE Confidence: 0.77793974
02:17:21.865 --> 02:17:23.226 at these stations because there's
NOTE Confidence: 0.77793974
02:17:23.226 --> 02:17:24.665 probably something that we haven't
NOTE Confidence: 0.77793974

02:17:24.665 --> 02:17:25.726 really appreciated.
NOTE Confidence: 0.83307093

02:17:26.266 --> 02:17:27.145 Should they send to them,
NOTE Confidence: 0.83307093

02:17:27.145 --> 02:17:28.426 I mean, it's valuable because
NOTE Confidence: 0.83307093

02:17:28.426 --> 02:17:29.625 it's so cost effective. They
NOTE Confidence: 0.83307093

02:17:29.625 --> 02:17:31.545 just buy it. Absolutely. And
NOTE Confidence: 0.83307093

02:17:31.545 --> 02:17:32.905 I would love to be
NOTE Confidence: 0.83307093

02:17:32.905 --> 02:17:34.045 in a project where
NOTE Confidence: 0.9558955

02:17:34.700 --> 02:17:35.900 we look at the genetic
NOTE Confidence: 0.9558955

02:17:35.900 --> 02:17:36.940 changes and then we go
NOTE Confidence: 0.9558955

02:17:36.940 --> 02:17:37.980 back to the tissues and
NOTE Confidence: 0.9558955

02:17:37.980 --> 02:17:39.420 say, okay. How how did
NOTE Confidence: 0.9558955

02:17:39.420 --> 02:17:40.720 this manifest histologically?
NOTE Confidence: 0.9858154

02:17:41.500 --> 02:17:42.940 That is being done in
NOTE Confidence: 0.9858154

02:17:42.940 --> 02:17:44.480 some places, but it's still
NOTE Confidence: 0.9180298

02:17:45.340 --> 02:17:47.120 early to say. Mina.
NOTE Confidence: 0.8886719

02:17:52.125 --> 02:17:52.625 Mina.

NOTE Confidence: 0.9301409
02:18:01.820 --> 02:18:02.900 I'm sorry. I couldn't hear
NOTE Confidence: 0.9301409
02:18:02.900 --> 02:18:03.681 you. Why?
NOTE Confidence: 0.7319336
02:18:06.940 --> 02:18:07.440 Oh.
NOTE Confidence: 0.9805814
02:18:09.740 --> 02:18:11.340 That's an interesting point. I
NOTE Confidence: 0.9805814
02:18:11.340 --> 02:18:12.700 don't know if we know
NOTE Confidence: 0.9805814
02:18:12.700 --> 02:18:14.320 exactly how the,
NOTE Confidence: 0.94323087
02:18:14.860 --> 02:18:16.645 effect of the diCircle one
NOTE Confidence: 0.94323087
02:18:16.645 --> 02:18:18.745 mutation leads to a cystic
NOTE Confidence: 0.94323087
02:18:18.805 --> 02:18:20.165 degeneration. I think that this
NOTE Confidence: 0.94323087
02:18:20.245 --> 02:18:21.545 well, the cystic formation.
NOTE Confidence: 0.9965515
02:18:22.004 --> 02:18:23.445 I think that the cystic
NOTE Confidence: 0.9965515
02:18:23.445 --> 02:18:25.285 formation may have more to
NOTE Confidence: 0.9965515
02:18:25.285 --> 02:18:27.705 do with the specific environment
NOTE Confidence: 0.9965515
02:18:27.765 --> 02:18:28.265 histologically
NOTE Confidence: 0.98779297
02:18:28.726 --> 02:18:29.625 that it happens
NOTE Confidence: 0.96157527

02:18:29.971 --> 02:18:31.171 In the lung, for example,
NOTE Confidence: 0.96157527

02:18:31.171 --> 02:18:33.171 is relatively easy to understand
NOTE Confidence: 0.96157527

02:18:33.171 --> 02:18:34.610 that there there is a
NOTE Confidence: 0.96157527

02:18:34.610 --> 02:18:36.311 flow of air around, etcetera.
NOTE Confidence: 0.96157527

02:18:36.610 --> 02:18:37.490 But I don't know in
NOTE Confidence: 0.96157527

02:18:37.490 --> 02:18:38.851 other places that are solid
NOTE Confidence: 0.96157527

02:18:38.851 --> 02:18:40.610 organs not exposed to these
NOTE Confidence: 0.96157527

02:18:40.610 --> 02:18:42.290 types of pressures. I really
NOTE Confidence: 0.96157527

02:18:42.290 --> 02:18:43.330 don't know. I don't know
NOTE Confidence: 0.96157527

02:18:43.330 --> 02:18:45.351 if anybody knows if,
NOTE Confidence: 0.6376953

02:18:57.985 --> 02:18:58.485 solid.
NOTE Confidence: 0.90352786

02:19:16.885 --> 02:19:18.084 In in the kidney too.
NOTE Confidence: 0.90352786

02:19:18.084 --> 02:19:19.705 In the kidney, cystic nephrona
NOTE Confidence: 0.9328613

02:19:20.084 --> 02:19:22.105 is, by definition, cystic.
NOTE Confidence: 0.68291557

02:19:22.565 --> 02:19:24.004 Yes. I think of the
NOTE Confidence: 0.68291557

02:19:24.004 --> 02:19:25.385 benign Yes. Yes. Yes.

NOTE Confidence: 0.22619629
02:19:41.351 --> 02:19:41.851 Express
NOTE Confidence: 0.91625977
02:19:42.615 --> 02:19:43.115 What,
NOTE Confidence: 0.79455566
02:19:43.655 --> 02:19:44.935 when, what time do you
NOTE Confidence: 0.79455566
02:19:44.935 --> 02:19:45.435 express
NOTE Confidence: 0.9161072
02:19:46.215 --> 02:19:47.595 a number of proteins?
NOTE Confidence: 0.99316406
02:19:47.975 --> 02:19:48.475 Right.
NOTE Confidence: 0.9873047
02:19:49.575 --> 02:19:50.075 And
NOTE Confidence: 0.8779297
02:19:50.455 --> 02:19:51.435 it is weird
NOTE Confidence: 0.96154785
02:19:51.895 --> 02:19:52.635 that epigenetics
NOTE Confidence: 0.7752228
02:20:11.254 --> 02:20:13.334 not just the sequence, but
NOTE Confidence: 0.7752228
02:20:13.334 --> 02:20:14.535 also some Epigenetics.
NOTE Confidence: 0.47888184
02:20:14.936 --> 02:20:15.436 Sequence,
NOTE Confidence: 0.782959
02:20:16.215 --> 02:20:16.715 like,
NOTE Confidence: 0.42749023
02:20:17.975 --> 02:20:18.475 upon,
NOTE Confidence: 0.8981323
02:20:19.175 --> 02:20:20.795 you know, that's Yep.
NOTE Confidence: 0.9587751

02:20:23.490 --> 02:20:24.770 I think that your comment
NOTE Confidence: 0.9587751

02:20:24.770 --> 02:20:25.430 is perfectly
NOTE Confidence: 0.7406006

02:20:25.890 --> 02:20:26.390 correct.
NOTE Confidence: 0.9614868

02:20:26.850 --> 02:20:28.609 And we are pushing for
NOTE Confidence: 0.9614868

02:20:28.770 --> 02:20:30.529 we as humans are pushing
NOTE Confidence: 0.9614868

02:20:30.529 --> 02:20:31.270 for that.
NOTE Confidence: 0.9980469

02:20:31.729 --> 02:20:32.229 However,
NOTE Confidence: 0.99934894

02:20:32.850 --> 02:20:33.830 it has had
NOTE Confidence: 1

02:20:34.290 --> 02:20:34.790 some
NOTE Confidence: 0.9759766

02:20:35.445 --> 02:20:37.705 undesirable effects in the community.
NOTE Confidence: 0.9849772

02:20:38.325 --> 02:20:39.686 One of the examples that
NOTE Confidence: 0.9849772

02:20:39.686 --> 02:20:40.965 I was discussing this morning
NOTE Confidence: 0.9849772

02:20:40.965 --> 02:20:42.745 with Anita Hudner was that
NOTE Confidence: 0.76000977

02:20:43.125 --> 02:20:44.584 the p, the CNS5
NOTE Confidence: 0.9696655

02:20:44.965 --> 02:20:46.265 edition of the WHO,
NOTE Confidence: 0.9505371

02:20:46.565 --> 02:20:48.325 the central nervous system tumor

NOTE Confidence: 0.9505371
02:20:48.325 --> 02:20:50.105 volume of the WHO incorporates
NOTE Confidence: 0.99886066
02:20:50.740 --> 02:20:53.080 a massive amount of sequencing
NOTE Confidence: 0.99886066
02:20:53.300 --> 02:20:53.800 and
NOTE Confidence: 0.92109376
02:20:54.180 --> 02:20:56.600 methylation, which is epigenetic changes,
NOTE Confidence: 0.8605957
02:20:56.979 --> 02:20:57.479 analysis.
NOTE Confidence: 0.947421
02:20:57.780 --> 02:20:59.620 And it incorporates them, many
NOTE Confidence: 0.947421
02:20:59.620 --> 02:21:01.700 of them as essential rather
NOTE Confidence: 0.947421
02:21:01.700 --> 02:21:02.200 than
NOTE Confidence: 0.98801
02:21:02.596 --> 02:21:04.436 desirable criteria. And the problem
NOTE Confidence: 0.98801
02:21:04.436 --> 02:21:06.056 with that is that
NOTE Confidence: 0.978125
02:21:06.516 --> 02:21:07.875 the over ninety percent of
NOTE Confidence: 0.978125
02:21:07.875 --> 02:21:09.315 the population in the world
NOTE Confidence: 0.978125
02:21:09.315 --> 02:21:10.855 has no access to that.
NOTE Confidence: 0.98950195
02:21:11.315 --> 02:21:13.415 So, yes, we need to
NOTE Confidence: 0.98950195
02:21:13.556 --> 02:21:14.216 be able
NOTE Confidence: 0.99902344

02:21:14.530 --> 02:21:17.030 to develop those approaches, those
NOTE Confidence: 0.99902344

02:21:17.250 --> 02:21:17.750 techniques
NOTE Confidence: 0.98657227

02:21:18.450 --> 02:21:20.390 to identify these changes
NOTE Confidence: 0.94710284

02:21:20.690 --> 02:21:22.770 but we cannot demand that
NOTE Confidence: 0.94710284

02:21:22.770 --> 02:21:23.270 everyone
NOTE Confidence: 0.99934894

02:21:23.890 --> 02:21:24.851 has to be able to
NOTE Confidence: 0.99934894

02:21:24.851 --> 02:21:25.351 diagnose
NOTE Confidence: 0.95362806

02:21:25.970 --> 02:21:27.774 a tumor with not only
NOTE Confidence: 0.95362806

02:21:27.774 --> 02:21:29.635 sequencing, but also with methylation
NOTE Confidence: 0.95362806

02:21:29.774 --> 02:21:31.615 analysis or other forms of
NOTE Confidence: 0.95362806

02:21:31.615 --> 02:21:33.535 epigenetic analysis. But I think
NOTE Confidence: 0.95362806

02:21:33.535 --> 02:21:34.975 that you are absolutely correct.
NOTE Confidence: 0.95362806

02:21:34.975 --> 02:21:36.015 This is a direction that
NOTE Confidence: 0.95362806

02:21:36.015 --> 02:21:36.834 we are following.
NOTE Confidence: 0.9937221

02:21:37.215 --> 02:21:37.935 I don't know if we
NOTE Confidence: 0.9937221

02:21:37.935 --> 02:21:39.535 will get there sooner or

NOTE Confidence: 0.9937221
02:21:39.535 --> 02:21:40.754 later, but we will.
NOTE Confidence: 0.73757327
02:21:41.891 --> 02:21:43.090 What is the focus of
NOTE Confidence: 0.73757327
02:21:43.090 --> 02:21:44.950 the non coding Jew? Right?
NOTE Confidence: 0.9666748
02:21:45.330 --> 02:21:46.391 Yeah. Exactly.
NOTE Confidence: 0.8000814
02:21:47.010 --> 02:21:48.770 Yeah. Junk. Yes. They they
NOTE Confidence: 0.8000814
02:21:48.770 --> 02:21:50.610 so called junk. Yeah. They
NOTE Confidence: 0.8000814
02:21:50.610 --> 02:21:52.311 hit the of chromaffin chromatin.
NOTE Confidence: 0.9301758
02:21:52.851 --> 02:21:53.351 Yes.
NOTE Confidence: 0.48597935
02:21:54.315 --> 02:21:55.436 Is it really a best
NOTE Confidence: 0.48597935
02:21:55.436 --> 02:21:56.095 in targeted
NOTE Confidence: 0.8186998
02:21:56.795 --> 02:21:58.314 therapy, you know, to prevent
NOTE Confidence: 0.8186998
02:21:58.314 --> 02:21:59.754 the tumor from alright. Once
NOTE Confidence: 0.8186998
02:21:59.754 --> 02:22:01.275 we know the nicer or
NOTE Confidence: 0.8186998
02:22:01.275 --> 02:22:03.035 then whatever genes that that
NOTE Confidence: 0.8186998
02:22:03.035 --> 02:22:04.555 we targeted to avoid the
NOTE Confidence: 0.8186998

02:22:04.555 --> 02:22:05.055 tumor
NOTE Confidence: 0.98480225

02:22:07.689 --> 02:22:08.970 Well, I think there is
NOTE Confidence: 0.98480225

02:22:08.970 --> 02:22:10.109 progress in terms
NOTE Confidence: 0.9946289

02:22:12.330 --> 02:22:13.069 of stopping
NOTE Confidence: 0.91831464

02:22:13.850 --> 02:22:15.609 the growth development in certain
NOTE Confidence: 0.91831464

02:22:15.609 --> 02:22:17.609 examples, but not in many
NOTE Confidence: 0.91831464

02:22:17.609 --> 02:22:18.515 examples. You
NOTE Confidence: 0.71435547

02:22:19.694 --> 02:22:20.194 you
NOTE Confidence: 0.88065964

02:22:20.495 --> 02:22:22.334 cannot change the genetic composition
NOTE Confidence: 0.88065964

02:22:22.334 --> 02:22:24.254 with targeted therapy. You can
NOTE Confidence: 0.88065964

02:22:24.254 --> 02:22:24.754 invalidate
NOTE Confidence: 0.8721369

02:22:25.375 --> 02:22:27.215 or annulate the effect of
NOTE Confidence: 0.8721369

02:22:27.215 --> 02:22:28.734 a given gene in in
NOTE Confidence: 0.8721369

02:22:28.734 --> 02:22:29.234 general.
NOTE Confidence: 0.8898926

02:22:29.935 --> 02:22:31.795 There is genetic therapy
NOTE Confidence: 0.9757857

02:22:32.420 --> 02:22:34.439 that is changing the genome

NOTE Confidence: 0.9757857

02:22:34.580 --> 02:22:36.520 in certain conditions, not necessarily

NOTE Confidence: 0.9757857

02:22:36.659 --> 02:22:37.159 neoplastic,

NOTE Confidence: 0.94492483

02:22:37.939 --> 02:22:40.100 Duchenne dystrophy, for example. There

NOTE Confidence: 0.94492483

02:22:40.100 --> 02:22:41.859 are examples of genetic approaches

NOTE Confidence: 0.94492483

02:22:41.859 --> 02:22:44.439 that correct the genetic abnormality

NOTE Confidence: 0.94492483

02:22:44.659 --> 02:22:46.279 and therefore will prevent

NOTE Confidence: 0.86150795

02:22:46.936 --> 02:22:49.275 the, continuation of the disorder.

NOTE Confidence: 0.86150795

02:22:49.334 --> 02:22:50.695 But Farzana may be able

NOTE Confidence: 0.86150795

02:22:50.695 --> 02:22:52.235 to respond to that question.

NOTE Confidence: 0.86150795

02:22:52.535 --> 02:22:53.735 Yeah. So I think the

NOTE Confidence: 0.86150795

02:22:53.815 --> 02:22:55.334 at the moment, these, like,

NOTE Confidence: 0.86150795

02:22:55.334 --> 02:22:57.254 diagnosis symptoms, etcetera, there is

NOTE Confidence: 0.86150795

02:22:57.254 --> 02:22:57.754 no

NOTE Confidence: 0.76464844

02:22:58.615 --> 02:22:59.895 talk with therapy. But I

NOTE Confidence: 0.76464844

02:22:59.895 --> 02:23:01.334 think the reason to diagnose

NOTE Confidence: 0.76464844

02:23:01.334 --> 02:23:02.075 them early
NOTE Confidence: 0.44975585

02:23:02.600 --> 02:23:04.220 is that we are screening.
NOTE Confidence: 0.72021484

02:23:05.000 --> 02:23:06.060 And we can actually
NOTE Confidence: 0.8087972

02:23:06.520 --> 02:23:08.119 the type three, for example,
NOTE Confidence: 0.8087972

02:23:08.119 --> 02:23:10.119 PPD is the individual treat.
NOTE Confidence: 0.8087972

02:23:10.119 --> 02:23:10.760 But if you have a
NOTE Confidence: 0.8087972

02:23:10.760 --> 02:23:12.279 type one heart, you can
NOTE Confidence: 0.8087972

02:23:12.279 --> 02:23:13.560 actually resect it and you're
NOTE Confidence: 0.8087972

02:23:13.560 --> 02:23:15.079 done. So I think early
NOTE Confidence: 0.8087972

02:23:15.079 --> 02:23:16.840 screening matches that. In fact,
NOTE Confidence: 0.8087972

02:23:16.840 --> 02:23:17.055 they
NOTE Confidence: 0.33735353

02:23:18.574 --> 02:23:19.955 were in in in in
NOTE Confidence: 0.77944803

02:23:20.334 --> 02:23:21.694 where the mother I mean,
NOTE Confidence: 0.77944803

02:23:21.694 --> 02:23:22.574 I just tell you an
NOTE Confidence: 0.77944803

02:23:22.574 --> 02:23:24.354 example of how screening helps.
NOTE Confidence: 0.77944803

02:23:24.495 --> 02:23:26.654 The mother had saw on

NOTE Confidence: 0.77944803
02:23:26.654 --> 02:23:27.154 CNN
NOTE Confidence: 0.7684733
02:23:27.854 --> 02:23:28.654 that if you have a
NOTE Confidence: 0.7684733
02:23:28.654 --> 02:23:29.154 sex
NOTE Confidence: 0.93359375
02:24:49.024 --> 02:24:49.604 I think
NOTE Confidence: 0.94731444
02:24:50.305 --> 02:24:51.524 I think it will happen,
NOTE Confidence: 0.99593097
02:24:51.984 --> 02:24:53.685 but it's not happening yet.
NOTE Confidence: 0.99593097
02:24:53.825 --> 02:24:54.325 Okay?
NOTE Confidence: 0.9638672
02:25:05.840 --> 02:25:07.840 Well, thank you again to
NOTE Confidence: 0.9638672
02:25:07.840 --> 02:25:08.979 all of you, and
NOTE Confidence: 0.9885254
02:25:09.840 --> 02:25:10.579 much appreciated.
NOTE Confidence: 0.19970703
02:26:27.320 --> 02:26:27.820 My
NOTE Confidence: 0.87597656
02:26:28.280 --> 02:26:28.780 computer.
NOTE Confidence: 0.80231935
02:26:32.601 --> 02:26:34.301 Okay. Can I leave now?
NOTE Confidence: 0.78137815
02:27:50.689 --> 02:27:51.970 Okay. Thank you. Thank you.
NOTE Confidence: 0.78137815
02:27:51.970 --> 02:27:53.590 Thank you. Okay. I am
NOTE Confidence: 0.9347957

02:27:53.970 --> 02:27:54.850 Rosie. Nice to meet you.

NOTE Confidence: 0.9347957

02:27:54.850 --> 02:27:55.970 Rosie, very nice to meet

NOTE Confidence: 0.9347957

02:27:55.970 --> 02:27:56.890 you. I'm Sarah.