

WEBVTT

NOTE duration: "00:33:13.216"

NOTE Confidence: 0.98745453

00:00:00.240 --> 00:00:01.439 My name is Rick Pierce,

NOTE Confidence: 0.98745453

00:00:01.439 --> 00:00:02.639 and I'm an associate professor

NOTE Confidence: 0.98745453

00:00:02.639 --> 00:00:04.000 of pediatrics in the section

NOTE Confidence: 0.98745453

00:00:04.000 --> 00:00:05.779 of pediatric critical care medicine

NOTE Confidence: 0.9921686

00:00:06.080 --> 00:00:07.359 at the Yale New Haven

NOTE Confidence: 0.9921686

00:00:07.359 --> 00:00:07.859 Children's

NOTE Confidence: 0.9550734

00:00:08.320 --> 00:00:10.000 Hospital and Yale School of

NOTE Confidence: 0.9550734

00:00:10.000 --> 00:00:10.500 Medicine.

NOTE Confidence: 0.9988527

00:00:10.880 --> 00:00:13.299 I'm talking today about pediatric

NOTE Confidence: 0.9972042

00:00:13.599 --> 00:00:15.540 systemic capillary leak syndrome

NOTE Confidence: 0.9501179

00:00:15.975 --> 00:00:17.175 and the challenges in its

NOTE Confidence: 0.9501179

00:00:17.175 --> 00:00:17.675 diagnosis

NOTE Confidence: 0.99902713

00:00:18.135 --> 00:00:18.875 and treatment.

NOTE Confidence: 0.99943745

00:00:19.335 --> 00:00:20.614 The purpose of this module

NOTE Confidence: 0.99943745

00:00:20.614 --> 00:00:22.235 is to raise awareness  
NOTE Confidence: 0.9994413

00:00:22.695 --> 00:00:23.675 about the diagnosis  
NOTE Confidence: 0.9654625

00:00:23.975 --> 00:00:26.154 and inpatient and outpatient treatment  
NOTE Confidence: 0.9654625

00:00:26.215 --> 00:00:28.154 of systemic capillary leak disease  
NOTE Confidence: 0.9654625

00:00:28.375 --> 00:00:29.095 so that it may be  
NOTE Confidence: 0.9654625

00:00:29.095 --> 00:00:29.915 better recognized  
NOTE Confidence: 0.99946404

00:00:30.375 --> 00:00:32.370 in critically ill children. Systemic  
NOTE Confidence: 0.99946404

00:00:32.370 --> 00:00:34.050 capillary leak disease is a  
NOTE Confidence: 0.99946404

00:00:34.050 --> 00:00:35.030 rare syndrome  
NOTE Confidence: 0.9990748

00:00:35.570 --> 00:00:37.890 characterized by episodes of recurrent  
NOTE Confidence: 0.9990748

00:00:37.890 --> 00:00:39.829 shock that are not related  
NOTE Confidence: 0.9990748

00:00:40.129 --> 00:00:41.430 to systemic infection.  
NOTE Confidence: 0.999799

00:00:42.050 --> 00:00:43.430 It's been called idiopathic  
NOTE Confidence: 0.9786431

00:00:43.985 --> 00:00:46.004 capillary leak syndrome or Clarkson's  
NOTE Confidence: 0.9786431

00:00:46.065 --> 00:00:47.425 disease because it was first  
NOTE Confidence: 0.9786431

00:00:47.425 --> 00:00:49.824 described by Bayard Clarkson in

NOTE Confidence: 0.9786431

00:00:49.824 --> 00:00:51.045 nineteen fifty eight.

NOTE Confidence: 0.9994291

00:00:51.504 --> 00:00:53.344 He described a woman who

NOTE Confidence: 0.9994291

00:00:53.344 --> 00:00:55.125 would have episodes of shock

NOTE Confidence: 0.9994291

00:00:55.344 --> 00:00:57.060 every five to eight months.

NOTE Confidence: 0.9547999

00:00:57.540 --> 00:00:58.660 She is shown here on

NOTE Confidence: 0.9547999

00:00:58.660 --> 00:00:59.700 the top picture in her

NOTE Confidence: 0.9547999

00:00:59.700 --> 00:01:01.300 well state and shown on

NOTE Confidence: 0.9547999

00:01:01.300 --> 00:01:03.060 the below in a state

NOTE Confidence: 0.9547999

00:01:03.060 --> 00:01:04.740 of leak where she has

NOTE Confidence: 0.9547999

00:01:04.740 --> 00:01:07.080 facial edema and extremity edema.

NOTE Confidence: 0.9493378

00:01:07.780 --> 00:01:08.840 She would have episodes,

NOTE Confidence: 0.9979655

00:01:09.515 --> 00:01:11.034 starting at thirty two years

NOTE Confidence: 0.9979655

00:01:11.034 --> 00:01:12.795 of age in nineteen fifty

NOTE Confidence: 0.9979655

00:01:12.795 --> 00:01:14.715 six. Her episodes would occur

NOTE Confidence: 0.9979655

00:01:14.715 --> 00:01:15.915 every five to eight months

NOTE Confidence: 0.9979655

00:01:15.915 --> 00:01:17.215 and progressively worsened.

NOTE Confidence: 0.99931043

00:01:17.595 --> 00:01:18.495 They were characterized

NOTE Confidence: 0.86362904

00:01:19.035 --> 00:01:19.535 by,

NOTE Confidence: 0.9990159

00:01:19.995 --> 00:01:20.495 malaise,

NOTE Confidence: 0.97983

00:01:21.275 --> 00:01:23.630 abdominal pain, nausea, and vomiting

NOTE Confidence: 0.97983

00:01:23.630 --> 00:01:24.990 for about thirty six hours,

NOTE Confidence: 0.97983

00:01:24.990 --> 00:01:26.110 and then she would have

NOTE Confidence: 0.97983

00:01:26.110 --> 00:01:27.250 extremity edema

NOTE Confidence: 0.99506825

00:01:27.950 --> 00:01:29.790 and hypotension. And her cases

NOTE Confidence: 0.99506825

00:01:29.790 --> 00:01:31.470 became her episodes of leak

NOTE Confidence: 0.99506825

00:01:31.470 --> 00:01:32.850 became worse and worse

NOTE Confidence: 0.7804978

00:01:33.150 --> 00:01:33.970 until unfortunately,

NOTE Confidence: 0.97421074

00:01:34.750 --> 00:01:36.110 she had an episode in

NOTE Confidence: 0.97421074

00:01:36.110 --> 00:01:37.605 nineteen fifty eight, which caused

NOTE Confidence: 0.97421074

00:01:37.725 --> 00:01:39.485 led to hypovolemic shock and

NOTE Confidence: 0.97421074

00:01:39.485 --> 00:01:41.745 caused a terminal cardiac arrest.

NOTE Confidence: 0.99465895

00:01:42.365 --> 00:01:43.425 Since that time,

NOTE Confidence: 0.9795134

00:01:43.885 --> 00:01:45.165 we've learned a lot about

NOTE Confidence: 0.9795134

00:01:45.165 --> 00:01:46.944 systemic capillary leak disease,

NOTE Confidence: 0.99007916

00:01:47.245 --> 00:01:49.165 and it's recharacterized by recurrent

NOTE Confidence: 0.99007916

00:01:49.165 --> 00:01:51.265 episodes of hypovolemic shock.

NOTE Confidence: 0.999736

00:01:51.770 --> 00:01:53.149 These episodes are recurrent

NOTE Confidence: 0.96922946

00:01:53.450 --> 00:01:55.390 and they're not explained by

NOTE Confidence: 0.96922946

00:01:55.450 --> 00:01:57.310 infection or other abnormalities.

NOTE Confidence: 0.999325

00:01:58.250 --> 00:02:00.729 These episodes typically have three

NOTE Confidence: 0.999325

00:02:00.729 --> 00:02:01.229 phases.

NOTE Confidence: 0.9993382

00:02:01.929 --> 00:02:03.450 The first phase is called

NOTE Confidence: 0.9993382

00:02:03.450 --> 00:02:04.189 the prodrome.

NOTE Confidence: 0.9934145

00:02:04.955 --> 00:02:06.655 This phase is quite nonspecific

NOTE Confidence: 0.9178831

00:02:07.195 --> 00:02:08.895 and can be composed

NOTE Confidence: 0.9178831

00:02:08.955 --> 00:02:10.175 of fever,

NOTE Confidence: 0.99970865

00:02:10.475 --> 00:02:11.455 general malaise,  
NOTE Confidence: 0.9997568

00:02:11.995 --> 00:02:13.215 abdominal pain,  
NOTE Confidence: 0.9533733

00:02:13.675 --> 00:02:14.175 myalgias,  
NOTE Confidence: 0.99064595

00:02:15.514 --> 00:02:17.115 and patients will typically have  
NOTE Confidence: 0.99064595

00:02:17.115 --> 00:02:19.190 lower urine output, be drinking  
NOTE Confidence: 0.99064595

00:02:19.190 --> 00:02:20.810 more, and have an increase  
NOTE Confidence: 0.99064595

00:02:21.030 --> 00:02:22.330 in body weight.  
NOTE Confidence: 0.9995585

00:02:22.630 --> 00:02:24.470 The prodrome phase may last  
NOTE Confidence: 0.9995585

00:02:24.470 --> 00:02:25.530 for several days  
NOTE Confidence: 0.9097902

00:02:25.910 --> 00:02:27.430 and then comes the leak  
NOTE Confidence: 0.9097902

00:02:27.430 --> 00:02:27.930 phase.  
NOTE Confidence: 0.9996012

00:02:28.709 --> 00:02:30.569 The leak phase is characterized  
NOTE Confidence: 0.99947006

00:02:30.870 --> 00:02:32.090 by hypotension,  
NOTE Confidence: 0.954184

00:02:32.790 --> 00:02:33.290 hypoalbuminemia,  
NOTE Confidence: 0.9800317

00:02:34.385 --> 00:02:35.204 and hemoconcentration.  
NOTE Confidence: 0.9994712

00:02:36.544 --> 00:02:38.864 These three findings constitute the

NOTE Confidence: 0.9994712  
00:02:38.864 --> 00:02:39.364 diagnostic  
NOTE Confidence: 0.9989348  
00:02:39.745 --> 00:02:41.984 triad of systemic capillary leak  
NOTE Confidence: 0.9989348  
00:02:41.984 --> 00:02:42.484 disease.  
NOTE Confidence: 0.9959575  
00:02:42.864 --> 00:02:44.064 Patients in the leak phase  
NOTE Confidence: 0.9959575  
00:02:44.064 --> 00:02:46.004 may also have generalized edema  
NOTE Confidence: 0.99369204  
00:02:46.305 --> 00:02:48.305 ranging from mild swelling of  
NOTE Confidence: 0.99369204  
00:02:48.305 --> 00:02:48.885 the extremities  
NOTE Confidence: 0.8806807  
00:02:49.530 --> 00:02:50.030 to,  
NOTE Confidence: 0.998757  
00:02:50.650 --> 00:02:51.790 severe ascites,  
NOTE Confidence: 0.9467062  
00:02:52.250 --> 00:02:53.150 pleural effusions,  
NOTE Confidence: 0.9997436  
00:02:53.530 --> 00:02:55.150 and even compartment syndrome  
NOTE Confidence: 0.9902221  
00:02:56.090 --> 00:02:57.389 of of the extremities.  
NOTE Confidence: 0.99912333  
00:02:57.930 --> 00:02:59.290 The leak phase may last  
NOTE Confidence: 0.99912333  
00:02:59.290 --> 00:03:00.989 for hours to days  
NOTE Confidence: 0.9766404  
00:03:01.305 --> 00:03:02.844 and then, transitions  
NOTE Confidence: 0.9975946

00:03:03.144 --> 00:03:04.525 rather suddenly and unexpectedly

NOTE Confidence: 0.99943656

00:03:04.984 --> 00:03:06.825 to the recovery phase. In

NOTE Confidence: 0.99943656

00:03:06.825 --> 00:03:08.745 the recovery phase, volume that

NOTE Confidence: 0.99943656

00:03:08.745 --> 00:03:09.704 has leaked out of the

NOTE Confidence: 0.99943656

00:03:09.704 --> 00:03:10.444 blood vessels

NOTE Confidence: 0.9780134

00:03:10.825 --> 00:03:12.745 is rapidly recruited back into

NOTE Confidence: 0.9780134

00:03:12.745 --> 00:03:14.370 the blood vessels and may

NOTE Confidence: 0.9780134

00:03:14.370 --> 00:03:16.549 result in sudden unexpected

NOTE Confidence: 0.9990728

00:03:16.849 --> 00:03:17.829 volume overload.

NOTE Confidence: 0.9996083

00:03:18.290 --> 00:03:19.269 This may produce

NOTE Confidence: 0.99956137

00:03:19.569 --> 00:03:20.629 pulmonary edema

NOTE Confidence: 0.99848014

00:03:20.930 --> 00:03:22.689 or cardiac failure from the

NOTE Confidence: 0.99848014

00:03:22.689 --> 00:03:24.709 sudden increase in intravascular

NOTE Confidence: 0.999856

00:03:25.090 --> 00:03:25.590 volume.

NOTE Confidence: 0.99272877

00:03:26.044 --> 00:03:27.565 The cause of SCLS is

NOTE Confidence: 0.99272877

00:03:27.565 --> 00:03:29.245 unknown, but there are two

NOTE Confidence: 0.99272877  
00:03:29.245 --> 00:03:29.745 general  
NOTE Confidence: 0.80455667  
00:03:30.285 --> 00:03:30.785 noncompeting  
NOTE Confidence: 0.9993936  
00:03:31.245 --> 00:03:31.745 theories.  
NOTE Confidence: 0.9355713  
00:03:32.044 --> 00:03:33.245 The first is that there  
NOTE Confidence: 0.9355713  
00:03:33.245 --> 00:03:34.144 is some systemic  
NOTE Confidence: 0.97137815  
00:03:34.845 --> 00:03:37.504 insult that results in a  
NOTE Confidence: 0.97137815  
00:03:37.565 --> 00:03:39.905 exaggerated response of pro inflammatory  
NOTE Confidence: 0.97137815  
00:03:40.125 --> 00:03:40.625 mediators.  
NOTE Confidence: 0.9991012  
00:03:41.280 --> 00:03:42.400 In this theory, the blood  
NOTE Confidence: 0.9991012  
00:03:42.400 --> 00:03:43.940 vessels are responding normally.  
NOTE Confidence: 0.97312206  
00:03:44.400 --> 00:03:45.920 The other theory is that  
NOTE Confidence: 0.97312206  
00:03:45.920 --> 00:03:47.780 there may be normal systemic  
NOTE Confidence: 0.97312206  
00:03:47.840 --> 00:03:49.940 responses to inflammation or insults  
NOTE Confidence: 0.97312206  
00:03:50.000 --> 00:03:51.060 and that the endothelial  
NOTE Confidence: 0.9996493  
00:03:51.440 --> 00:03:51.940 cells  
NOTE Confidence: 0.91400975

00:03:52.240 --> 00:03:54.260 are hyper reactive to normal  
NOTE Confidence: 0.91400975  
00:03:54.400 --> 00:03:54.900 stimuli.  
NOTE Confidence: 0.99944913  
00:03:55.655 --> 00:03:57.275 Both theories have been supported  
NOTE Confidence: 0.99448305  
00:03:57.655 --> 00:03:59.435 by circumstantial evidence.  
NOTE Confidence: 0.9739165  
00:04:00.055 --> 00:04:02.315 Supporting the circulating pro inflammatory  
NOTE Confidence: 0.9739165  
00:04:02.535 --> 00:04:04.535 mediator theory are studies that  
NOTE Confidence: 0.9739165  
00:04:04.535 --> 00:04:06.375 have shown that patients have  
NOTE Confidence: 0.9739165  
00:04:06.375 --> 00:04:07.435 abnormal paraproteins.  
NOTE Confidence: 0.99638206  
00:04:08.200 --> 00:04:08.939 These are  
NOTE Confidence: 0.92786306  
00:04:09.480 --> 00:04:10.459 these are termed  
NOTE Confidence: 0.99724203  
00:04:10.760 --> 00:04:11.579 the monoclonal  
NOTE Confidence: 0.95119816  
00:04:11.959 --> 00:04:14.760 gammopathy of undetermined significance that  
NOTE Confidence: 0.95119816  
00:04:14.760 --> 00:04:15.900 we'll talk about later,  
NOTE Confidence: 0.949066  
00:04:16.520 --> 00:04:19.320 or vascular targeted cytokines such  
NOTE Confidence: 0.949066  
00:04:19.320 --> 00:04:20.620 as vascular endothelial  
NOTE Confidence: 0.95855165  
00:04:21.000 --> 00:04:22.380 growth factor or VEGF,

NOTE Confidence: 0.9860725  
00:04:22.865 --> 00:04:25.285 interleukin two, angiopoietin two,  
NOTE Confidence: 0.90857285  
00:04:25.825 --> 00:04:26.325 leukotrienes,  
NOTE Confidence: 0.99914116  
00:04:26.785 --> 00:04:28.325 or other chemokines  
NOTE Confidence: 0.98504794  
00:04:28.785 --> 00:04:29.765 or cytokines.  
NOTE Confidence: 0.998079  
00:04:30.305 --> 00:04:31.825 These levels may be elevated  
NOTE Confidence: 0.998079  
00:04:31.825 --> 00:04:33.185 in patients when they're sick  
NOTE Confidence: 0.998079  
00:04:33.185 --> 00:04:34.560 compared to when they're well  
NOTE Confidence: 0.998079  
00:04:34.960 --> 00:04:36.720 or elevated in patients with  
NOTE Confidence: 0.998079  
00:04:36.720 --> 00:04:38.419 capillary leak disease compared  
NOTE Confidence: 0.9702831  
00:04:38.800 --> 00:04:40.320 to not. But they're hard  
NOTE Confidence: 0.9702831  
00:04:40.320 --> 00:04:41.919 to interpret because of all  
NOTE Confidence: 0.9702831  
00:04:41.919 --> 00:04:43.940 of those multiple comparisons.  
NOTE Confidence: 0.99933803  
00:04:44.400 --> 00:04:46.100 And no studies have conclusively  
NOTE Confidence: 0.99933803  
00:04:46.320 --> 00:04:48.525 shown a single systemic  
NOTE Confidence: 0.9990528  
00:04:48.904 --> 00:04:51.085 cytokine or chemokine is responsible  
NOTE Confidence: 0.9975436

00:04:51.705 --> 00:04:53.305 for the leak phase of  
NOTE Confidence: 0.9975436

00:04:53.305 --> 00:04:55.005 systemic capillary leak disease.  
NOTE Confidence: 0.9837576

00:04:55.945 --> 00:04:57.225 The other theory is that  
NOTE Confidence: 0.9837576

00:04:57.225 --> 00:04:59.005 the endothelial cells are hyperreactive,  
NOTE Confidence: 0.9994377

00:04:59.625 --> 00:05:01.330 and this theory is supported  
NOTE Confidence: 0.9992401

00:05:01.789 --> 00:05:03.889 by increased markers of endothelial  
NOTE Confidence: 0.9992401

00:05:04.029 --> 00:05:05.009 cell apoptosis  
NOTE Confidence: 0.9842092

00:05:05.629 --> 00:05:07.150 in patients during the leak  
NOTE Confidence: 0.9842092

00:05:07.150 --> 00:05:08.449 phase or  
NOTE Confidence: 0.99851894

00:05:08.750 --> 00:05:10.509 isolation of endothelial cells from  
NOTE Confidence: 0.99851894

00:05:10.509 --> 00:05:11.569 patients with SCLS  
NOTE Confidence: 0.87478405

00:05:12.029 --> 00:05:13.810 and investigating their overreactive  
NOTE Confidence: 0.9874682

00:05:14.270 --> 00:05:16.215 signaling in the lab in  
NOTE Confidence: 0.9874682

00:05:16.215 --> 00:05:17.355 response to,  
NOTE Confidence: 0.99861526

00:05:17.895 --> 00:05:19.515 normal circulating cytokines.  
NOTE Confidence: 0.96167916

00:05:20.455 --> 00:05:21.975 And this is what, I

NOTE Confidence: 0.96167916  
00:05:21.975 --> 00:05:23.335 do in addition to caring  
NOTE Confidence: 0.96167916  
00:05:23.335 --> 00:05:24.935 for critically ill children is  
NOTE Confidence: 0.96167916  
00:05:24.935 --> 00:05:27.335 I run a, research lab  
NOTE Confidence: 0.96167916  
00:05:27.335 --> 00:05:27.835 focused  
NOTE Confidence: 0.9807184  
00:05:28.139 --> 00:05:30.220 on blood vessel dysfunction in  
NOTE Confidence: 0.9807184  
00:05:30.220 --> 00:05:31.360 critically ill children.  
NOTE Confidence: 0.9988567  
00:05:31.740 --> 00:05:33.440 And I am keenly interested  
NOTE Confidence: 0.9998193  
00:05:33.900 --> 00:05:34.800 in understanding  
NOTE Confidence: 0.982658  
00:05:35.339 --> 00:05:36.779 how the junctions in blood  
NOTE Confidence: 0.982658  
00:05:36.779 --> 00:05:38.860 vessels break down and allow  
NOTE Confidence: 0.982658  
00:05:38.860 --> 00:05:40.960 leak from inside the vascular  
NOTE Confidence: 0.982658  
00:05:41.100 --> 00:05:42.620 space to outside of the  
NOTE Confidence: 0.982658  
00:05:42.620 --> 00:05:43.680 vascular space.  
NOTE Confidence: 0.9802145  
00:05:44.205 --> 00:05:45.565 The goals of my laboratory  
NOTE Confidence: 0.9802145  
00:05:45.565 --> 00:05:46.765 are to understand this, how  
NOTE Confidence: 0.9802145

00:05:46.765 --> 00:05:48.045 this happens in critically ill  
NOTE Confidence: 0.9802145

00:05:48.045 --> 00:05:50.605 children, and identify ways to  
NOTE Confidence: 0.9802145

00:05:50.605 --> 00:05:53.005 ultimately treat it to reduce  
NOTE Confidence: 0.9802145

00:05:53.005 --> 00:05:54.685 the burden of critical illness  
NOTE Confidence: 0.9802145

00:05:54.685 --> 00:05:55.425 in children.  
NOTE Confidence: 0.98852986

00:05:56.220 --> 00:05:58.060 I do research patients with  
NOTE Confidence: 0.98852986

00:05:58.060 --> 00:05:59.979 systemic capillary leak disease, and  
NOTE Confidence: 0.98852986

00:05:59.979 --> 00:06:01.020 my goal is to understand  
NOTE Confidence: 0.98852986

00:06:01.020 --> 00:06:02.940 how vascular leak occurs in  
NOTE Confidence: 0.98852986

00:06:02.940 --> 00:06:04.460 those patients so that we  
NOTE Confidence: 0.98852986

00:06:04.460 --> 00:06:05.819 may develop new treatments for  
NOTE Confidence: 0.98852986

00:06:05.819 --> 00:06:07.580 them, but also a better  
NOTE Confidence: 0.98852986

00:06:07.580 --> 00:06:09.680 understand how vascular leak occurs  
NOTE Confidence: 0.98852986

00:06:09.819 --> 00:06:11.520 in all critically ill children  
NOTE Confidence: 0.98852986

00:06:11.794 --> 00:06:12.855 and develop therapies  
NOTE Confidence: 0.99776506

00:06:13.315 --> 00:06:14.995 to reduce that leak and

NOTE Confidence: 0.99776506  
00:06:14.995 --> 00:06:16.435 reduce the time children have  
NOTE Confidence: 0.99776506  
00:06:16.435 --> 00:06:17.635 to stay in the intensive  
NOTE Confidence: 0.99776506  
00:06:17.635 --> 00:06:18.375 care unit.  
NOTE Confidence: 0.9882775  
00:06:19.635 --> 00:06:21.014 The first case of pediatric  
NOTE Confidence: 0.9882775  
00:06:21.154 --> 00:06:23.235 capillary leak disease was reported  
NOTE Confidence: 0.9882775  
00:06:23.235 --> 00:06:24.675 in nineteen ninety five. This  
NOTE Confidence: 0.9882775  
00:06:24.675 --> 00:06:25.794 was a nine year old  
NOTE Confidence: 0.9882775  
00:06:25.794 --> 00:06:26.935 Canadian female,  
NOTE Confidence: 0.9967904  
00:06:27.520 --> 00:06:29.379 and she started having episodes  
NOTE Confidence: 0.9967904  
00:06:29.439 --> 00:06:30.960 at age three. And she  
NOTE Confidence: 0.9967904  
00:06:30.960 --> 00:06:33.039 had episodes every eight to  
NOTE Confidence: 0.9967904  
00:06:33.039 --> 00:06:33.860 nine months,  
NOTE Confidence: 0.97764724  
00:06:34.479 --> 00:06:34.979 until,  
NOTE Confidence: 0.9713466  
00:06:36.080 --> 00:06:37.919 she became almost nine years  
NOTE Confidence: 0.9713466  
00:06:37.919 --> 00:06:39.120 old when she had an  
NOTE Confidence: 0.9713466

00:06:39.120 --> 00:06:41.315 episode that again resulted in  
NOTE Confidence: 0.9713466

00:06:41.315 --> 00:06:41.815 hypovolemic  
NOTE Confidence: 0.99907464

00:06:42.195 --> 00:06:44.595 shock, cardiac arrest, and her  
NOTE Confidence: 0.99907464

00:06:44.595 --> 00:06:45.815 unfortunate death.  
NOTE Confidence: 0.9991058

00:06:46.355 --> 00:06:48.035 The youngest patient described with  
NOTE Confidence: 0.9991058

00:06:48.035 --> 00:06:50.275 capillary leak disease was a  
NOTE Confidence: 0.9991058

00:06:50.275 --> 00:06:52.215 twenty three week premature infant  
NOTE Confidence: 0.99460065

00:06:52.600 --> 00:06:54.360 who developed their first episode  
NOTE Confidence: 0.99460065

00:06:54.360 --> 00:06:55.560 of leak at twenty eight  
NOTE Confidence: 0.99460065

00:06:55.560 --> 00:06:57.000 days of life or a  
NOTE Confidence: 0.99460065

00:06:57.000 --> 00:06:59.400 corrected gestational age of twenty  
NOTE Confidence: 0.99460065

00:06:59.400 --> 00:07:01.900 seven weeks. So systemic capillary  
NOTE Confidence: 0.99460065

00:07:01.960 --> 00:07:03.720 leak disease has been reported  
NOTE Confidence: 0.99460065

00:07:03.720 --> 00:07:04.940 in very young babies,  
NOTE Confidence: 0.9998362

00:07:05.720 --> 00:07:06.779 very young children,  
NOTE Confidence: 0.9364155

00:07:07.755 --> 00:07:08.255 adolescents,

NOTE Confidence: 0.996619

00:07:08.714 --> 00:07:10.095 and now adults.

NOTE Confidence: 0.9900965

00:07:11.035 --> 00:07:12.955 Since nineteen ninety five, there

NOTE Confidence: 0.9900965

00:07:12.955 --> 00:07:14.415 are about thirty cases

NOTE Confidence: 0.9845671

00:07:14.794 --> 00:07:16.955 of pediatric systemic capillary leak

NOTE Confidence: 0.9845671

00:07:16.955 --> 00:07:17.455 identified,

NOTE Confidence: 0.99815536

00:07:17.835 --> 00:07:19.035 and we've not been able

NOTE Confidence: 0.99815536

00:07:19.035 --> 00:07:20.880 to determine any sex,

NOTE Confidence: 0.9721002

00:07:21.280 --> 00:07:23.520 ethnicity, or inheritance patterns for

NOTE Confidence: 0.9721002

00:07:23.520 --> 00:07:24.180 this disease,

NOTE Confidence: 0.9829378

00:07:24.480 --> 00:07:26.480 although that remains an area

NOTE Confidence: 0.9829378

00:07:26.480 --> 00:07:28.480 of ongoing research. About three

NOTE Confidence: 0.9829378

00:07:28.480 --> 00:07:30.020 hundred adults have been identified

NOTE Confidence: 0.9829378

00:07:30.080 --> 00:07:31.520 with this disease, and children

NOTE Confidence: 0.9829378

00:07:31.520 --> 00:07:33.600 have the same disease phases

NOTE Confidence: 0.9829378

00:07:33.600 --> 00:07:34.975 as adults. That is they

NOTE Confidence: 0.9829378

00:07:34.975 --> 00:07:36.514 have this prodrome  
NOTE Confidence: 0.99033755

00:07:36.895 --> 00:07:38.815 of not feeling well, a  
NOTE Confidence: 0.99033755

00:07:38.815 --> 00:07:40.815 sudden leak phase followed by  
NOTE Confidence: 0.99033755

00:07:40.815 --> 00:07:42.895 a sudden recovery phase. But  
NOTE Confidence: 0.99033755

00:07:42.895 --> 00:07:44.435 there are reasons to think  
NOTE Confidence: 0.99033755

00:07:44.495 --> 00:07:46.815 that pediatric systemic capillary leak  
NOTE Confidence: 0.99033755

00:07:46.815 --> 00:07:48.595 disease may be different from  
NOTE Confidence: 0.99033755

00:07:48.815 --> 00:07:50.960 adult systemic capillary leak disease.  
NOTE Confidence: 0.99033755

00:07:51.260 --> 00:07:52.880 And those differences are summarized  
NOTE Confidence: 0.99033755

00:07:53.020 --> 00:07:54.480 in, this table.  
NOTE Confidence: 0.98626757

00:07:55.020 --> 00:07:55.520 First,  
NOTE Confidence: 0.9967118

00:07:56.460 --> 00:07:58.080 are the demographic data.  
NOTE Confidence: 0.98816

00:07:58.620 --> 00:07:59.980 Although more than three hundred  
NOTE Confidence: 0.98816

00:07:59.980 --> 00:08:01.580 adults with SCLS have been  
NOTE Confidence: 0.98816

00:08:01.580 --> 00:08:03.420 described, only about thirty children  
NOTE Confidence: 0.98816

00:08:03.420 --> 00:08:05.200 with SCLS have been described.

NOTE Confidence: 0.99027574

00:08:05.634 --> 00:08:06.995 And the average age of

NOTE Confidence: 0.99027574

00:08:06.995 --> 00:08:08.294 diagnosis is about,

NOTE Confidence: 0.9908127

00:08:08.754 --> 00:08:10.275 five point five years or

NOTE Confidence: 0.9908127

00:08:10.275 --> 00:08:11.094 young childhood

NOTE Confidence: 0.989938

00:08:11.395 --> 00:08:12.995 in children, whereas adults are

NOTE Confidence: 0.989938

00:08:12.995 --> 00:08:14.675 diagnosed into middle age, about

NOTE Confidence: 0.989938

00:08:14.675 --> 00:08:16.055 forty two years of age.

NOTE Confidence: 0.989938

00:08:16.354 --> 00:08:18.134 Children also much more frequently

NOTE Confidence: 0.989938

00:08:18.275 --> 00:08:19.974 have a viral prodrome

NOTE Confidence: 0.77506495

00:08:20.275 --> 00:08:20.935 that is

NOTE Confidence: 0.9956445

00:08:21.370 --> 00:08:23.710 fever, abdominal pain, myalgias,

NOTE Confidence: 0.98911357

00:08:24.410 --> 00:08:26.090 as opposed to adult capillary

NOTE Confidence: 0.98911357

00:08:26.090 --> 00:08:27.530 leak disease where about fifty

NOTE Confidence: 0.98911357

00:08:27.530 --> 00:08:29.069 percent of patients have those.

NOTE Confidence: 0.97473395

00:08:29.610 --> 00:08:31.210 And then another striking difference

NOTE Confidence: 0.97473395

00:08:31.210 --> 00:08:33.370 is the monoclonal gammopathy of

NOTE Confidence: 0.97473395

00:08:33.370 --> 00:08:33.870 undetermined

NOTE Confidence: 0.9614192

00:08:43.675 --> 00:08:44.175 SCLS.

NOTE Confidence: 0.992337

00:08:44.554 --> 00:08:45.595 So this is a very

NOTE Confidence: 0.992337

00:08:45.595 --> 00:08:46.875 big difference and may tell

NOTE Confidence: 0.992337

00:08:46.875 --> 00:08:48.075 us that there's some difference

NOTE Confidence: 0.992337

00:08:48.075 --> 00:08:49.640 in the underlying pathology,

NOTE Confidence: 0.92760074

00:08:50.020 --> 00:08:51.700 but we haven't determined what

NOTE Confidence: 0.92760074

00:08:51.700 --> 00:08:52.680 this means yet.

NOTE Confidence: 0.9543585

00:08:53.220 --> 00:08:55.300 Finally, the time between episodes

NOTE Confidence: 0.9543585

00:08:55.300 --> 00:08:56.120 and diagnoses

NOTE Confidence: 0.9995834

00:08:56.500 --> 00:08:57.800 is more than a year

NOTE Confidence: 0.9603577

00:08:58.180 --> 00:08:59.860 in adult SCLS and we

NOTE Confidence: 0.9603577

00:08:59.860 --> 00:09:01.860 don't know that interval in

NOTE Confidence: 0.9603577

00:09:01.860 --> 00:09:03.105 pediatric SCLS.

NOTE Confidence: 0.97916657

00:09:03.485 --> 00:09:05.085 And we also don't know

NOTE Confidence: 0.97916657  
00:09:05.085 --> 00:09:06.385 the five year mortality,  
NOTE Confidence: 0.9994555  
00:09:06.845 --> 00:09:07.345 which  
NOTE Confidence: 0.9847805  
00:09:08.205 --> 00:09:10.445 untreated was about seventy five  
NOTE Confidence: 0.9847805  
00:09:10.445 --> 00:09:12.045 percent in adults, but newer  
NOTE Confidence: 0.9847805  
00:09:12.045 --> 00:09:12.545 treatments  
NOTE Confidence: 0.9866937  
00:09:12.925 --> 00:09:14.304 that I'll talk about later  
NOTE Confidence: 0.9866937  
00:09:14.525 --> 00:09:15.425 have dramatically  
NOTE Confidence: 0.9683621  
00:09:15.965 --> 00:09:16.380 reduced  
NOTE Confidence: 0.9901481  
00:09:17.340 --> 00:09:18.880 that. But these are unknown  
NOTE Confidence: 0.9901481  
00:09:18.940 --> 00:09:20.320 in children with SCLS  
NOTE Confidence: 0.9219946  
00:09:21.020 --> 00:09:22.700 and many children may have  
NOTE Confidence: 0.9219946  
00:09:22.700 --> 00:09:23.679 several episodes  
NOTE Confidence: 0.94605017  
00:09:24.140 --> 00:09:26.400 before capillary leak is diagnosed.  
NOTE Confidence: 0.96354985  
00:09:26.780 --> 00:09:28.620 And these episodes may be  
NOTE Confidence: 0.96354985  
00:09:28.620 --> 00:09:29.120 severe,  
NOTE Confidence: 0.9994059

00:09:29.580 --> 00:09:30.385 which is one of the  
NOTE Confidence: 0.9994059

00:09:30.385 --> 00:09:32.005 major points of this education  
NOTE Confidence: 0.9994059

00:09:32.065 --> 00:09:32.565 module  
NOTE Confidence: 0.97230303

00:09:32.945 --> 00:09:35.025 is that pediatric systemic capillary  
NOTE Confidence: 0.97230303

00:09:35.025 --> 00:09:36.885 leak disease is likely underdiagnosed  
NOTE Confidence: 0.9991356

00:09:37.985 --> 00:09:39.345 due to a lack of  
NOTE Confidence: 0.9991356

00:09:39.345 --> 00:09:40.565 provider awareness  
NOTE Confidence: 0.9957676

00:09:40.945 --> 00:09:41.925 about the disease.  
NOTE Confidence: 0.9917093

00:09:42.510 --> 00:09:43.710 And this is very, very  
NOTE Confidence: 0.9917093

00:09:43.710 --> 00:09:45.309 important because these episodes can  
NOTE Confidence: 0.9917093

00:09:45.309 --> 00:09:46.130 be severe.  
NOTE Confidence: 0.9990339

00:09:46.670 --> 00:09:48.290 They can be severe hypovolemic  
NOTE Confidence: 0.99557185

00:09:48.670 --> 00:09:50.350 shock. They can have severe  
NOTE Confidence: 0.99557185

00:09:50.350 --> 00:09:51.790 problems, but they can also  
NOTE Confidence: 0.99557185

00:09:51.790 --> 00:09:52.610 be prevented  
NOTE Confidence: 0.9983998

00:09:53.230 --> 00:09:54.190 if we can make the

NOTE Confidence: 0.9983998  
00:09:54.190 --> 00:09:54.690 diagnosis  
NOTE Confidence: 0.9997878  
00:09:55.309 --> 00:09:55.809 early.  
NOTE Confidence: 0.996691  
00:09:56.315 --> 00:09:57.355 I'd like to talk about  
NOTE Confidence: 0.996691  
00:09:57.355 --> 00:09:59.595 a typical case presentation for  
NOTE Confidence: 0.996691  
00:09:59.595 --> 00:10:02.175 pediatric systemic capillary leak disease  
NOTE Confidence: 0.996691  
00:10:02.475 --> 00:10:04.735 and include a parent testimonial  
NOTE Confidence: 0.996691  
00:10:04.955 --> 00:10:06.075 that is used with their  
NOTE Confidence: 0.996691  
00:10:06.075 --> 00:10:06.575 consent.  
NOTE Confidence: 0.97768825  
00:10:07.355 --> 00:10:08.795 Children typically will present to  
NOTE Confidence: 0.97768825  
00:10:08.795 --> 00:10:10.735 the emergency room with edema  
NOTE Confidence: 0.97768825  
00:10:10.795 --> 00:10:12.380 and distributive shock.  
NOTE Confidence: 0.99974215  
00:10:12.760 --> 00:10:14.220 Key elements of the history  
NOTE Confidence: 0.99426144  
00:10:14.600 --> 00:10:16.140 include the patient was otherwise  
NOTE Confidence: 0.99426144  
00:10:16.200 --> 00:10:18.220 healthy. They may have a  
NOTE Confidence: 0.99426144  
00:10:18.360 --> 00:10:20.360 vague viral prodrome for two  
NOTE Confidence: 0.99426144

00:10:20.360 --> 00:10:21.640 to three days that include  
NOTE Confidence: 0.99426144

00:10:21.640 --> 00:10:23.900 things like abdominal pain,  
NOTE Confidence: 0.94906753

00:10:25.035 --> 00:10:25.995 a little bit of eating  
NOTE Confidence: 0.94906753

00:10:25.995 --> 00:10:26.954 less, a little bit of  
NOTE Confidence: 0.94906753

00:10:26.954 --> 00:10:27.454 nausea,  
NOTE Confidence: 0.99693567

00:10:28.954 --> 00:10:29.454 myalgias,  
NOTE Confidence: 0.96662396

00:10:29.915 --> 00:10:31.834 low grade fever, but non  
NOTE Confidence: 0.96662396

00:10:32.154 --> 00:10:32.654 nonspecific  
NOTE Confidence: 0.9994872

00:10:33.194 --> 00:10:34.415 prodromal symptoms.  
NOTE Confidence: 0.97005934

00:10:35.194 --> 00:10:35.995 They may have had a  
NOTE Confidence: 0.97005934

00:10:35.995 --> 00:10:37.134 history of unexplained,  
NOTE Confidence: 0.9874988

00:10:37.675 --> 00:10:39.855 episodes of facial or extremity  
NOTE Confidence: 0.9874988

00:10:39.995 --> 00:10:40.495 edema,  
NOTE Confidence: 0.9996983

00:10:41.090 --> 00:10:42.929 and there's an absence of  
NOTE Confidence: 0.9996983

00:10:42.929 --> 00:10:43.830 known allergies  
NOTE Confidence: 0.9833701

00:10:44.290 --> 00:10:46.130 or other triggering factors such

NOTE Confidence: 0.9833701

00:10:46.130 --> 00:10:48.710 as drugs, infection, environmental exposures,

NOTE Confidence: 0.99973196

00:10:49.170 --> 00:10:50.710 or other chronic diseases

NOTE Confidence: 0.9891625

00:10:51.170 --> 00:10:53.030 that would suggest or or

NOTE Confidence: 0.9891625

00:10:53.330 --> 00:10:54.070 lead to,

NOTE Confidence: 0.9997136

00:10:54.530 --> 00:10:55.030 edema.

NOTE Confidence: 0.99619234

00:10:56.195 --> 00:10:57.235 Some key elements of the

NOTE Confidence: 0.99619234

00:10:57.235 --> 00:10:59.155 physical exam include those that

NOTE Confidence: 0.99619234

00:10:59.155 --> 00:11:01.415 are consistent with distributive shock.

NOTE Confidence: 0.99619234

00:11:01.475 --> 00:11:02.595 These patients will have a

NOTE Confidence: 0.99619234

00:11:02.595 --> 00:11:03.715 high heart rate and a

NOTE Confidence: 0.99619234

00:11:03.715 --> 00:11:04.775 low blood pressure.

NOTE Confidence: 0.9241765

00:11:05.235 --> 00:11:06.515 They may have,

NOTE Confidence: 0.99633

00:11:07.395 --> 00:11:10.054 signs of volume overload already

NOTE Confidence: 0.99633

00:11:10.340 --> 00:11:11.540 such as a third heart

NOTE Confidence: 0.99633

00:11:11.540 --> 00:11:13.059 sound or crackles on their

NOTE Confidence: 0.99633

00:11:13.059 --> 00:11:13.880 lung exam.  
NOTE Confidence: 0.995608

00:11:14.420 --> 00:11:16.200 Their skin color is typically,  
NOTE Confidence: 0.9658657

00:11:17.140 --> 00:11:19.480 modeled consistent with distributive shock,  
NOTE Confidence: 0.9658657

00:11:19.620 --> 00:11:21.080 and their temperature and capillary  
NOTE Confidence: 0.9658657

00:11:21.140 --> 00:11:22.900 refill time may also be  
NOTE Confidence: 0.9658657

00:11:22.900 --> 00:11:23.400 abnormal.  
NOTE Confidence: 0.99938464

00:11:24.035 --> 00:11:25.635 These patients usually do not  
NOTE Confidence: 0.99938464

00:11:25.635 --> 00:11:26.775 have a rash.  
NOTE Confidence: 0.9691034

00:11:27.155 --> 00:11:28.515 Their mental status may be  
NOTE Confidence: 0.9691034

00:11:28.515 --> 00:11:30.035 decreased, and if available at  
NOTE Confidence: 0.9691034

00:11:30.035 --> 00:11:30.775 your institution,  
NOTE Confidence: 0.96068347

00:11:31.235 --> 00:11:32.434 they may have you may  
NOTE Confidence: 0.96068347

00:11:32.434 --> 00:11:33.395 be able to do point  
NOTE Confidence: 0.96068347

00:11:33.395 --> 00:11:35.075 of care heart or lung  
NOTE Confidence: 0.96068347

00:11:35.075 --> 00:11:36.215 ultrasound revealing  
NOTE Confidence: 0.95532894

00:11:37.155 --> 00:11:39.095 increased, cardiac function,

NOTE Confidence: 0.9873891

00:11:39.650 --> 00:11:40.950 pericardial effusions,

NOTE Confidence: 0.90582335

00:11:41.570 --> 00:11:44.390 and plural effusions or pulmonary

NOTE Confidence: 0.90582335

00:11:44.450 --> 00:11:46.710 parenchymal edema by lung ultrasound.

NOTE Confidence: 0.9647771

00:11:47.170 --> 00:11:47.670 Reverie,

NOTE Confidence: 0.9992864

00:11:48.690 --> 00:11:50.370 woke up in April of

NOTE Confidence: 0.9992864

00:11:50.370 --> 00:11:51.590 twenty twenty two

NOTE Confidence: 0.9911815

00:11:52.285 --> 00:11:55.005 with very swollen eyes. So

NOTE Confidence: 0.9911815

00:11:55.005 --> 00:11:56.065 it was bilateral

NOTE Confidence: 0.97486633

00:11:56.445 --> 00:11:57.565 swelling in both of her

NOTE Confidence: 0.97486633

00:11:57.565 --> 00:11:59.265 eyes, periorbital edema,

NOTE Confidence: 0.95940757

00:11:59.645 --> 00:12:00.765 which we now know is

NOTE Confidence: 0.95940757

00:12:00.765 --> 00:12:01.905 the official term.

NOTE Confidence: 0.9985592

00:12:02.765 --> 00:12:04.125 And we thought it was

NOTE Confidence: 0.9985592

00:12:04.125 --> 00:12:04.625 allergies.

NOTE Confidence: 0.9953755

00:12:05.965 --> 00:12:07.240 At that time, she was

NOTE Confidence: 0.9953755

00:12:07.240 --> 00:12:08.540 not acting lethargic  
NOTE Confidence: 0.99520093

00:12:09.320 --> 00:12:11.100 or ill in any way.  
NOTE Confidence: 0.9609631

00:12:11.720 --> 00:12:13.899 We phoned the doctor and  
NOTE Confidence: 0.990483

00:12:14.360 --> 00:12:15.880 were instructed to start a  
NOTE Confidence: 0.990483

00:12:15.880 --> 00:12:17.420 typical course of antihistamines.  
NOTE Confidence: 0.95624983

00:12:18.040 --> 00:12:19.320 In that first episode, no  
NOTE Confidence: 0.95624983

00:12:19.320 --> 00:12:20.679 fever, nothing like that. We  
NOTE Confidence: 0.95624983

00:12:20.679 --> 00:12:22.254 thought, oh, freak incident. And  
NOTE Confidence: 0.95624983

00:12:22.254 --> 00:12:23.774 then lo and behold, come  
NOTE Confidence: 0.95624983

00:12:23.774 --> 00:12:25.535 July, she's running a fever  
NOTE Confidence: 0.95624983

00:12:25.535 --> 00:12:26.995 and her eyes start swelling.  
NOTE Confidence: 0.95624983

00:12:27.214 --> 00:12:28.415 And that's where we really  
NOTE Confidence: 0.95624983

00:12:28.415 --> 00:12:29.535 were like, this is not  
NOTE Confidence: 0.95624983

00:12:29.535 --> 00:12:31.295 allergies. Something else is going  
NOTE Confidence: 0.95624983

00:12:31.295 --> 00:12:31.795 on.  
NOTE Confidence: 0.9839846

00:12:33.134 --> 00:12:35.475 Second time, they said, okay.

NOTE Confidence: 0.9839846  
00:12:35.774 --> 00:12:37.225 Let's put a referral into,  
NOTE Confidence: 0.9617783  
00:12:38.070 --> 00:12:38.730 eye doctor.  
NOTE Confidence: 0.9857788  
00:12:39.510 --> 00:12:40.230 So then we did a  
NOTE Confidence: 0.9857788  
00:12:40.230 --> 00:12:41.590 referral to eye doctor. Eye  
NOTE Confidence: 0.9857788  
00:12:41.590 --> 00:12:43.030 doctor did not do tests,  
NOTE Confidence: 0.9857788  
00:12:43.030 --> 00:12:44.070 but they did do a  
NOTE Confidence: 0.9857788  
00:12:44.070 --> 00:12:45.910 full full exam. It was  
NOTE Confidence: 0.9857788  
00:12:45.910 --> 00:12:47.210 a pediatric ophthalmologist,  
NOTE Confidence: 0.9754837  
00:12:47.830 --> 00:12:48.809 not an optometrist.  
NOTE Confidence: 0.98183733  
00:12:49.590 --> 00:12:50.710 So that was her second  
NOTE Confidence: 0.98183733  
00:12:50.710 --> 00:12:51.165 week.  
NOTE Confidence: 0.9979228  
00:12:51.964 --> 00:12:53.425 When that came back clear,  
NOTE Confidence: 0.9979228  
00:12:53.565 --> 00:12:54.304 the ophthalmologist  
NOTE Confidence: 0.9465498  
00:12:55.245 --> 00:12:56.545 sent us to the ENT.  
NOTE Confidence: 0.97368187  
00:12:57.404 --> 00:12:59.184 The ENT did do testing  
NOTE Confidence: 0.97368187

00:12:59.245 --> 00:13:00.605 day of. They did that  
NOTE Confidence: 0.97368187

00:13:00.605 --> 00:13:01.904 c the head CT,  
NOTE Confidence: 0.9976531

00:13:03.404 --> 00:13:04.144 but insurance  
NOTE Confidence: 0.9885321

00:13:05.490 --> 00:13:06.770 requires them to do, like,  
NOTE Confidence: 0.9885321

00:13:06.770 --> 00:13:08.610 thirty days of antibiotics, which  
NOTE Confidence: 0.9885321

00:13:08.610 --> 00:13:09.810 I'm sure you guys have  
NOTE Confidence: 0.9885321

00:13:09.810 --> 00:13:12.050 heard with yep. So she  
NOTE Confidence: 0.9885321

00:13:12.130 --> 00:13:13.170 they put her on thirty  
NOTE Confidence: 0.9885321

00:13:13.170 --> 00:13:15.190 days of antibiotics for a  
NOTE Confidence: 0.9885321

00:13:15.410 --> 00:13:17.490 potential what if sinus infection  
NOTE Confidence: 0.9885321

00:13:17.490 --> 00:13:18.895 that's, like, stuck somewhere in  
NOTE Confidence: 0.9885321

00:13:18.895 --> 00:13:20.735 her system. Third and fourth  
NOTE Confidence: 0.9885321

00:13:20.735 --> 00:13:22.095 one, we went to urgent  
NOTE Confidence: 0.9885321

00:13:22.095 --> 00:13:22.995 care twice,  
NOTE Confidence: 0.9541777

00:13:23.855 --> 00:13:25.395 and they only did urine  
NOTE Confidence: 0.9541777

00:13:25.455 --> 00:13:25.955 protein,

NOTE Confidence: 0.99041224  
00:13:26.495 --> 00:13:28.515 zero labs, no labs.  
NOTE Confidence: 0.99062294  
00:13:29.695 --> 00:13:30.195 And  
NOTE Confidence: 0.91954947  
00:13:30.575 --> 00:13:32.015 like I said, her fourth  
NOTE Confidence: 0.91954947  
00:13:32.015 --> 00:13:32.515 one,  
NOTE Confidence: 0.95251966  
00:13:33.300 --> 00:13:35.059 the which was influenza a  
NOTE Confidence: 0.95251966  
00:13:35.059 --> 00:13:37.300 October twenty two, she  
NOTE Confidence: 0.95251966  
00:13:37.300 --> 00:13:39.140 was very sick, and we  
NOTE Confidence: 0.95251966  
00:13:39.220 --> 00:13:41.059 I'm upset that there was  
NOTE Confidence: 0.95251966  
00:13:41.059 --> 00:13:43.320 no further care administered.  
NOTE Confidence: 0.9525216  
00:13:43.779 --> 00:13:45.300 She was, like, two weeks  
NOTE Confidence: 0.9525216  
00:13:45.300 --> 00:13:47.059 of school for sure. And  
NOTE Confidence: 0.9525216  
00:13:47.059 --> 00:13:48.420 she was febrile for  
NOTE Confidence: 0.9935559  
00:13:49.154 --> 00:13:50.514 I feel like her body  
NOTE Confidence: 0.9935559  
00:13:50.514 --> 00:13:51.954 temperature did not get back  
NOTE Confidence: 0.9935559  
00:13:51.954 --> 00:13:53.235 to, like, ninety eight six  
NOTE Confidence: 0.9935559

00:13:53.235 --> 00:13:54.434 for, like, a month. And  
NOTE Confidence: 0.9935559

00:13:54.434 --> 00:13:56.134 for that swelling to resolve,  
NOTE Confidence: 0.9935559

00:13:56.195 --> 00:13:57.475 it was every bit of  
NOTE Confidence: 0.9935559

00:13:57.475 --> 00:13:58.134 two weeks.  
NOTE Confidence: 0.98864925

00:13:58.595 --> 00:14:01.394 Yeah. In May of twenty  
NOTE Confidence: 0.98864925

00:14:01.394 --> 00:14:02.995 twenty three, it was Cinco  
NOTE Confidence: 0.98864925

00:14:02.995 --> 00:14:03.975 de Mayo weekend.  
NOTE Confidence: 0.96407825

00:14:04.670 --> 00:14:07.009 And once again, she was  
NOTE Confidence: 0.96407825

00:14:07.069 --> 00:14:09.889 very, very ill, fever, lethargic,  
NOTE Confidence: 0.99215597

00:14:10.589 --> 00:14:11.389 and it got to the  
NOTE Confidence: 0.99215597

00:14:11.389 --> 00:14:12.670 point where she was unable  
NOTE Confidence: 0.99215597

00:14:12.670 --> 00:14:14.050 to walk. And  
NOTE Confidence: 0.94356316

00:14:14.750 --> 00:14:16.690 that's when the red flags,  
NOTE Confidence: 0.99590576

00:14:17.035 --> 00:14:17.915 I was like, this is  
NOTE Confidence: 0.99590576

00:14:17.915 --> 00:14:19.115 just not right. She was  
NOTE Confidence: 0.99590576

00:14:19.115 --> 00:14:20.975 very slow to respond verbally.

NOTE Confidence: 0.99937165

00:14:21.435 --> 00:14:22.395 As you can see from

NOTE Confidence: 0.99937165

00:14:22.395 --> 00:14:23.455 the patient testimonial,

NOTE Confidence: 0.99982

00:14:24.235 --> 00:14:25.595 the signs and symptoms may

NOTE Confidence: 0.99982

00:14:25.595 --> 00:14:26.575 not be conclusive

NOTE Confidence: 0.9745628

00:14:27.035 --> 00:14:28.015 of any diagnosis.

NOTE Confidence: 0.96021855

00:14:28.970 --> 00:14:30.430 And the acute presentation,

NOTE Confidence: 0.9663912

00:14:30.970 --> 00:14:32.670 severity of shock, and availability

NOTE Confidence: 0.9663912

00:14:32.890 --> 00:14:34.570 of life saving personnel and

NOTE Confidence: 0.9663912

00:14:34.570 --> 00:14:35.070 resources

NOTE Confidence: 0.9807592

00:14:35.530 --> 00:14:37.290 may limit the consideration of

NOTE Confidence: 0.9807592

00:14:37.290 --> 00:14:39.150 a thorough differential diagnosis.

NOTE Confidence: 0.9941518

00:14:39.930 --> 00:14:42.430 But not all distributive shock

NOTE Confidence: 0.9941102

00:14:42.834 --> 00:14:44.455 is sepsis, and it's important

NOTE Confidence: 0.9941102

00:14:44.515 --> 00:14:46.535 to have a broader differential

NOTE Confidence: 0.9857621

00:14:47.075 --> 00:14:49.315 when considering patients who have

NOTE Confidence: 0.9857621

00:14:49.315 --> 00:14:50.995 come in one or more  
NOTE Confidence: 0.9857621

00:14:50.995 --> 00:14:53.154 time with the somewhat unique  
NOTE Confidence: 0.9857621

00:14:53.154 --> 00:14:54.214 finding of  
NOTE Confidence: 0.99950194

00:14:54.834 --> 00:14:55.334 hypotension  
NOTE Confidence: 0.8776319

00:14:55.795 --> 00:14:57.015 distributive shock  
NOTE Confidence: 0.9999143

00:14:57.330 --> 00:14:57.830 with  
NOTE Confidence: 0.9975169

00:14:58.210 --> 00:15:00.210 edema. The differential is listed  
NOTE Confidence: 0.9975169

00:15:00.210 --> 00:15:01.170 here on the left side  
NOTE Confidence: 0.9975169

00:15:01.170 --> 00:15:01.990 of this table.  
NOTE Confidence: 0.9656704

00:15:02.450 --> 00:15:04.130 Systemic capillary leak disease is  
NOTE Confidence: 0.9656704

00:15:04.130 --> 00:15:06.130 on top, and sepsis, the  
NOTE Confidence: 0.9656704

00:15:06.130 --> 00:15:08.370 most common etiology confused with  
NOTE Confidence: 0.9656704

00:15:08.370 --> 00:15:10.050 systemic capillary leak disease, is  
NOTE Confidence: 0.9656704

00:15:10.050 --> 00:15:11.190 on the next line.  
NOTE Confidence: 0.97233075

00:15:11.535 --> 00:15:13.135 Other specific causes of sepsis  
NOTE Confidence: 0.97233075

00:15:13.135 --> 00:15:14.675 such as toxic shock syndrome

NOTE Confidence: 0.97233075

00:15:14.894 --> 00:15:16.595 or inborn errors of immunity

NOTE Confidence: 0.97233075

00:15:16.654 --> 00:15:17.714 that lead to,

NOTE Confidence: 0.9721181

00:15:18.255 --> 00:15:19.855 shock states are shown there

NOTE Confidence: 0.9721181

00:15:19.855 --> 00:15:21.555 as well, along with anaphylaxis,

NOTE Confidence: 0.99311554

00:15:22.014 --> 00:15:24.514 nephrotic syndrome, hereditary angioedema,

NOTE Confidence: 0.9942195

00:15:25.600 --> 00:15:26.820 and Addison's disease.

NOTE Confidence: 0.9991915

00:15:27.280 --> 00:15:28.880 These differentials may seem broad,

NOTE Confidence: 0.9991915

00:15:28.880 --> 00:15:30.560 but you'll see how certain

NOTE Confidence: 0.9991915

00:15:30.560 --> 00:15:32.660 parts of different diseases overlap

NOTE Confidence: 0.9871581

00:15:32.960 --> 00:15:34.880 with different parts of the

NOTE Confidence: 0.9871581

00:15:34.880 --> 00:15:37.460 systemic capillary leak disease history,

NOTE Confidence: 0.9871581

00:15:37.520 --> 00:15:40.020 physical exam, clinical course, laboratory

NOTE Confidence: 0.9871581

00:15:40.080 --> 00:15:40.580 findings,

NOTE Confidence: 0.998049

00:15:40.935 --> 00:15:42.135 and how they may be

NOTE Confidence: 0.998049

00:15:42.135 --> 00:15:44.375 confused for systemic capillary leak

NOTE Confidence: 0.998049

00:15:44.375 --> 00:15:44.875 disease.  
NOTE Confidence: 0.975262

00:15:45.575 --> 00:15:47.015 Discussing the history and physical  
NOTE Confidence: 0.975262

00:15:47.015 --> 00:15:47.515 exam,  
NOTE Confidence: 0.94290406

00:15:48.135 --> 00:15:50.215 there in systemic capillary leak  
NOTE Confidence: 0.94290406

00:15:50.215 --> 00:15:51.654 disease, there may be one  
NOTE Confidence: 0.94290406

00:15:51.654 --> 00:15:52.955 or more of these episodes,  
NOTE Confidence: 0.9136364

00:15:53.895 --> 00:15:56.100 wet patients coming in with  
NOTE Confidence: 0.9136364

00:15:56.259 --> 00:15:58.519 the unique combination of hypotension  
NOTE Confidence: 0.9136364

00:15:58.819 --> 00:15:59.720 and edema.  
NOTE Confidence: 0.9786697

00:16:00.339 --> 00:16:02.100 In septic shock, toxic shock  
NOTE Confidence: 0.9786697

00:16:02.100 --> 00:16:03.940 syndrome, or septic shock from  
NOTE Confidence: 0.9786697

00:16:03.940 --> 00:16:05.319 inborn errors of immunity,  
NOTE Confidence: 0.9903089

00:16:05.779 --> 00:16:07.060 patients may come in with  
NOTE Confidence: 0.9903089

00:16:07.060 --> 00:16:09.079 fever and hypotension and tachycardia  
NOTE Confidence: 0.9428621

00:16:09.459 --> 00:16:11.220 very similar to systemic capillary  
NOTE Confidence: 0.9428621

00:16:11.220 --> 00:16:11.959 leak disease,

NOTE Confidence: 0.99368995

00:16:12.475 --> 00:16:15.055 but the signs of edema

NOTE Confidence: 0.99368995

00:16:15.195 --> 00:16:16.095 will be absent,

NOTE Confidence: 0.9820039

00:16:16.555 --> 00:16:17.835 and they may have other

NOTE Confidence: 0.9820039

00:16:17.835 --> 00:16:19.935 findings such as rash

NOTE Confidence: 0.9834768

00:16:20.635 --> 00:16:22.335 or a known genetic abnormality.

NOTE Confidence: 0.99956477

00:16:23.195 --> 00:16:24.875 Patients with anaphylaxis may come

NOTE Confidence: 0.99956477

00:16:24.875 --> 00:16:25.695 in with hypotension

NOTE Confidence: 0.8667979

00:16:26.395 --> 00:16:26.895 and,

NOTE Confidence: 0.99064225

00:16:27.540 --> 00:16:29.800 some edema. Typically, that's localized

NOTE Confidence: 0.99064225

00:16:29.860 --> 00:16:30.740 to the face or to

NOTE Confidence: 0.99064225

00:16:30.740 --> 00:16:31.779 the area where there was

NOTE Confidence: 0.99064225

00:16:31.779 --> 00:16:33.220 an exposure to a known

NOTE Confidence: 0.99064225

00:16:33.220 --> 00:16:33.720 allergen.

NOTE Confidence: 0.97435105

00:16:34.100 --> 00:16:35.779 These patients will also have

NOTE Confidence: 0.97435105

00:16:35.779 --> 00:16:38.180 profound respiratory distress such as,

NOTE Confidence: 0.97435105

00:16:38.340 --> 00:16:40.279 manifested by wheezing,  
NOTE Confidence: 0.99981

00:16:40.980 --> 00:16:42.360 or airway edema.  
NOTE Confidence: 0.99703807

00:16:43.005 --> 00:16:44.365 They may also have a  
NOTE Confidence: 0.99703807

00:16:44.365 --> 00:16:45.665 urticarial rash.  
NOTE Confidence: 0.99574965

00:16:46.445 --> 00:16:48.845 Patients with hereditary angioedema may  
NOTE Confidence: 0.99574965

00:16:48.845 --> 00:16:50.925 also have localized edema to  
NOTE Confidence: 0.99574965

00:16:50.925 --> 00:16:52.525 the face. They typically will  
NOTE Confidence: 0.99574965

00:16:52.525 --> 00:16:53.825 come in not with cardiovascular  
NOTE Confidence: 0.9886709

00:16:54.205 --> 00:16:55.885 symptoms, but with signs of  
NOTE Confidence: 0.9886709

00:16:55.885 --> 00:16:57.965 respiratory distress from upper airway  
NOTE Confidence: 0.9886709

00:16:57.965 --> 00:16:58.465 swelling.  
NOTE Confidence: 0.9711566

00:16:59.180 --> 00:17:00.800 Patients with nephrotic syndrome,  
NOTE Confidence: 0.9635075

00:17:01.180 --> 00:17:02.380 may come in with edema,  
NOTE Confidence: 0.9635075

00:17:02.380 --> 00:17:03.820 but typically these patients are  
NOTE Confidence: 0.9635075

00:17:03.820 --> 00:17:05.580 hypertensive and their course has  
NOTE Confidence: 0.9635075

00:17:05.580 --> 00:17:07.200 been more gradual onset.

NOTE Confidence: 0.997359

00:17:07.980 --> 00:17:09.740 And patients with Addison's disease

NOTE Confidence: 0.997359

00:17:09.740 --> 00:17:10.940 may also have a gradual

NOTE Confidence: 0.997359

00:17:10.940 --> 00:17:12.059 onset and come in with

NOTE Confidence: 0.997359

00:17:12.059 --> 00:17:13.580 hypotension, but there may be

NOTE Confidence: 0.997359

00:17:13.580 --> 00:17:15.544 some skin pigmentation findings.

NOTE Confidence: 0.9901575

00:17:16.484 --> 00:17:17.705 So what are the initial

NOTE Confidence: 0.9901575

00:17:17.845 --> 00:17:19.445 workup, for a patient with

NOTE Confidence: 0.9901575

00:17:19.445 --> 00:17:21.445 systemic capillary leak disease? Well,

NOTE Confidence: 0.9901575

00:17:21.445 --> 00:17:23.125 the initial focus has to

NOTE Confidence: 0.9901575

00:17:23.125 --> 00:17:25.044 be on the distributive shock.

NOTE Confidence: 0.9901575

00:17:25.044 --> 00:17:26.005 And so you would do

NOTE Confidence: 0.9901575

00:17:26.005 --> 00:17:27.044 all of the things at

NOTE Confidence: 0.9901575

00:17:27.044 --> 00:17:27.784 your institution

NOTE Confidence: 0.99975497

00:17:28.590 --> 00:17:30.990 associated with your septic shock

NOTE Confidence: 0.99975497

00:17:30.990 --> 00:17:31.490 bundle

NOTE Confidence: 0.98809844

00:17:32.190 --> 00:17:33.470 or, that you would do  
NOTE Confidence: 0.98809844

00:17:33.470 --> 00:17:35.390 for distributive shock. These may  
NOTE Confidence: 0.98809844

00:17:35.390 --> 00:17:36.850 include blood cultures, urinalysis,  
NOTE Confidence: 0.97235763

00:17:37.150 --> 00:17:39.410 urine cultures, CBC, complete metabolic  
NOTE Confidence: 0.97235763

00:17:39.470 --> 00:17:41.490 panel. Depending on the severity  
NOTE Confidence: 0.996263

00:17:42.305 --> 00:17:43.365 of the presentation,  
NOTE Confidence: 0.9370204

00:17:43.905 --> 00:17:44.785 you you may get a  
NOTE Confidence: 0.9370204

00:17:44.785 --> 00:17:45.744 blood gas to look at  
NOTE Confidence: 0.9370204

00:17:45.744 --> 00:17:47.425 gas exchange or perfusion with  
NOTE Confidence: 0.9370204

00:17:47.425 --> 00:17:47.925 lactate.  
NOTE Confidence: 0.983136

00:17:48.305 --> 00:17:49.665 If you have suspicion of  
NOTE Confidence: 0.983136

00:17:49.665 --> 00:17:51.585 decreased cardiac output, you may  
NOTE Confidence: 0.983136

00:17:51.585 --> 00:17:53.525 consider cardiac enzymes or natriuretic  
NOTE Confidence: 0.983136

00:17:53.665 --> 00:17:54.165 peptides,  
NOTE Confidence: 0.933829

00:17:54.830 --> 00:17:56.609 a tox screen, and coagulation  
NOTE Confidence: 0.933829

00:17:56.830 --> 00:17:57.330 panel.

NOTE Confidence: 0.9779384

00:17:58.270 --> 00:17:59.710 When these, come back, a

NOTE Confidence: 0.9779384

00:17:59.710 --> 00:18:01.950 secondary evaluation may include chest

NOTE Confidence: 0.9779384

00:18:01.950 --> 00:18:02.450 radiography

NOTE Confidence: 0.93976563

00:18:02.750 --> 00:18:04.510 or CT scan to look

NOTE Confidence: 0.93976563

00:18:04.510 --> 00:18:05.810 for a nidus of infection,

NOTE Confidence: 0.9642847

00:18:06.350 --> 00:18:07.570 other viral testing,

NOTE Confidence: 0.9876833

00:18:07.965 --> 00:18:09.184 or more,

NOTE Confidence: 0.96949995

00:18:09.885 --> 00:18:11.885 in in-depth testing such as

NOTE Confidence: 0.96949995

00:18:11.885 --> 00:18:14.044 cortisol, thyroid, or cytokine panels

NOTE Confidence: 0.96949995

00:18:14.044 --> 00:18:15.484 as they're available at your

NOTE Confidence: 0.96949995

00:18:15.484 --> 00:18:15.984 institution.

NOTE Confidence: 0.9691732

00:18:16.445 --> 00:18:17.325 We took her to the

NOTE Confidence: 0.9691732

00:18:17.325 --> 00:18:19.325 ED and, of course, she

NOTE Confidence: 0.9691732

00:18:19.325 --> 00:18:21.005 presented with a very high

NOTE Confidence: 0.9691732

00:18:21.005 --> 00:18:23.050 heart rate. So they immediately

NOTE Confidence: 0.9691732

00:18:23.190 --> 00:18:24.090 started fluids,  
NOTE Confidence: 0.9560806

00:18:25.030 --> 00:18:26.470 which, of course, is a  
NOTE Confidence: 0.9560806

00:18:26.470 --> 00:18:27.690 detrimental error  
NOTE Confidence: 0.978307

00:18:28.070 --> 00:18:29.190 and a reason that a  
NOTE Confidence: 0.978307

00:18:29.190 --> 00:18:30.330 lot of these kiddos  
NOTE Confidence: 0.99635935

00:18:30.710 --> 00:18:32.710 end up in ICU type  
NOTE Confidence: 0.99635935

00:18:32.710 --> 00:18:33.210 situations.  
NOTE Confidence: 0.9456938

00:18:33.670 --> 00:18:35.450 Her lower extremity swelled.  
NOTE Confidence: 0.9408075

00:18:37.154 --> 00:18:38.434 You started to see just  
NOTE Confidence: 0.9408075

00:18:38.434 --> 00:18:40.195 overall puffiness and Yes. To  
NOTE Confidence: 0.9408075

00:18:40.195 --> 00:18:40.914 the point where you couldn't  
NOTE Confidence: 0.9408075

00:18:40.914 --> 00:18:41.955 pull her shorts up over  
NOTE Confidence: 0.9408075

00:18:41.955 --> 00:18:43.154 her legs. Yes. And, you  
NOTE Confidence: 0.9408075

00:18:43.154 --> 00:18:44.034 know, you pick her up.  
NOTE Confidence: 0.9408075

00:18:44.034 --> 00:18:45.075 And and at that point,  
NOTE Confidence: 0.9408075

00:18:45.075 --> 00:18:45.734 she weighed,

NOTE Confidence: 0.99253243  
00:18:47.234 --> 00:18:48.994 I think, forty pounds or  
NOTE Confidence: 0.99253243  
00:18:48.994 --> 00:18:50.274 so, and she gained five  
NOTE Confidence: 0.99253243  
00:18:50.274 --> 00:18:51.930 pounds overnight. I know with  
NOTE Confidence: 0.99253243  
00:18:52.010 --> 00:18:53.369 other kids who ended up  
NOTE Confidence: 0.99253243  
00:18:53.369 --> 00:18:54.970 in the ICU, they were  
NOTE Confidence: 0.99253243  
00:18:54.970 --> 00:18:56.410 treating for what they thought  
NOTE Confidence: 0.99253243  
00:18:56.410 --> 00:18:58.350 was sepsis or septic shock,  
NOTE Confidence: 0.9355921  
00:18:58.970 --> 00:19:00.430 hence the fluid overload.  
NOTE Confidence: 0.99486333  
00:19:01.610 --> 00:19:03.070 Once we got her labs  
NOTE Confidence: 0.99486333  
00:19:03.210 --> 00:19:04.810 back with low albumin and  
NOTE Confidence: 0.99486333  
00:19:04.810 --> 00:19:05.630 low sodium,  
NOTE Confidence: 0.96023  
00:19:06.274 --> 00:19:07.955 we insisted on being transferred  
NOTE Confidence: 0.96023  
00:19:07.955 --> 00:19:09.015 to Big Duke.  
NOTE Confidence: 0.9987224  
00:19:09.394 --> 00:19:11.575 The laboratory findings of SCLS  
NOTE Confidence: 0.9998394  
00:19:12.115 --> 00:19:13.734 are quite unique as well.  
NOTE Confidence: 0.9983249

00:19:14.434 --> 00:19:16.195 Again, the diagnostic triad of  
NOTE Confidence: 0.9983249

00:19:16.195 --> 00:19:17.575 this is hypotension,  
NOTE Confidence: 0.97696096

00:19:18.310 --> 00:19:19.590 which can be as severe  
NOTE Confidence: 0.97696096

00:19:19.590 --> 00:19:21.750 as hypotensive shock and cardiac  
NOTE Confidence: 0.97696096

00:19:21.750 --> 00:19:22.250 arrest,  
NOTE Confidence: 0.93124914

00:19:22.550 --> 00:19:23.050 hemoconcentration,  
NOTE Confidence: 0.89333427

00:19:24.150 --> 00:19:24.810 where patients  
NOTE Confidence: 0.98518026

00:19:25.590 --> 00:19:27.750 hematocrit may double or even  
NOTE Confidence: 0.98518026

00:19:27.750 --> 00:19:29.530 triple from their baseline levels,  
NOTE Confidence: 0.98002696

00:19:30.070 --> 00:19:30.970 and hypoalbuminemia,  
NOTE Confidence: 0.9425033

00:19:32.284 --> 00:19:34.365 where unreadably low albumin is  
NOTE Confidence: 0.9425033

00:19:34.365 --> 00:19:34.865 common,  
NOTE Confidence: 0.9922747

00:19:35.244 --> 00:19:36.625 but at least below,  
NOTE Confidence: 0.9970139

00:19:37.085 --> 00:19:38.125 one and a half or  
NOTE Confidence: 0.9970139

00:19:38.125 --> 00:19:40.065 two milligrams per deciliter.  
NOTE Confidence: 0.9858775

00:19:40.525 --> 00:19:41.725 These patients will also have

NOTE Confidence: 0.9858775

00:19:41.725 --> 00:19:43.825 high lactate, but normal inflammatory

NOTE Confidence: 0.9711043

00:19:44.125 --> 00:19:46.330 markers, which will differentiate them

NOTE Confidence: 0.9711043

00:19:46.330 --> 00:19:48.730 from sepsis, toxic shock syndrome,

NOTE Confidence: 0.9711043

00:19:48.730 --> 00:19:50.490 and many inborn errors of

NOTE Confidence: 0.9711043

00:19:50.490 --> 00:19:50.990 immunity.

NOTE Confidence: 0.9479614

00:19:51.770 --> 00:19:53.530 Other tests that may clue

NOTE Confidence: 0.9479614

00:19:53.530 --> 00:19:54.809 into a diet alternate

NOTE Confidence: 0.9750497

00:19:55.210 --> 00:19:56.490 other tests that may clue

NOTE Confidence: 0.9750497

00:19:56.490 --> 00:19:57.950 into an alternate diagnosis

NOTE Confidence: 0.97944677

00:19:58.625 --> 00:20:01.125 include elevated tryptase for anaphylaxis,

NOTE Confidence: 0.9719412

00:20:02.304 --> 00:20:04.884 abnormal complement levels for hereditary

NOTE Confidence: 0.97042525

00:20:05.184 --> 00:20:05.684 angioedema,

NOTE Confidence: 0.9988791

00:20:06.465 --> 00:20:08.484 and abnormal lipid profiles

NOTE Confidence: 0.9779873

00:20:08.865 --> 00:20:10.244 for nephrotic syndrome,

NOTE Confidence: 0.9983411

00:20:10.945 --> 00:20:12.404 and low cortisol

NOTE Confidence: 0.9409577

00:20:12.705 --> 00:20:13.205 or  
NOTE Confidence: 0.9949385

00:20:13.859 --> 00:20:14.359 ACTH  
NOTE Confidence: 0.99273735

00:20:14.980 --> 00:20:16.440 for Addison's disease.  
NOTE Confidence: 0.99979895

00:20:18.020 --> 00:20:19.000 The initial treatment  
NOTE Confidence: 0.99253124

00:20:19.300 --> 00:20:22.100 for pediatric SCLS disease has  
NOTE Confidence: 0.99253124

00:20:22.100 --> 00:20:23.220 to be focused on the  
NOTE Confidence: 0.99253124

00:20:23.220 --> 00:20:25.400 severity of shock. This includes  
NOTE Confidence: 0.99253124

00:20:25.460 --> 00:20:27.060 stabilizing the airway and breathing  
NOTE Confidence: 0.99253124

00:20:27.060 --> 00:20:28.359 with supplemental oxygen  
NOTE Confidence: 0.998851

00:20:28.865 --> 00:20:31.125 or mechanical support as needed.  
NOTE Confidence: 0.9979151

00:20:31.585 --> 00:20:33.105 Next, you must stabilize the  
NOTE Confidence: 0.9979151

00:20:33.105 --> 00:20:33.605 cardiovascular  
NOTE Confidence: 0.89297074

00:20:34.225 --> 00:20:34.725 dysfunction,  
NOTE Confidence: 0.9986749

00:20:35.185 --> 00:20:36.465 and this is done with  
NOTE Confidence: 0.9986749

00:20:36.465 --> 00:20:37.205 a conservative  
NOTE Confidence: 0.9990916

00:20:37.744 --> 00:20:38.565 fluid resuscitation

NOTE Confidence: 0.9992371

00:20:39.025 --> 00:20:41.345 strategy. Patients with systemic capillary

NOTE Confidence: 0.9992371

00:20:41.345 --> 00:20:42.565 leak disease are

NOTE Confidence: 0.9991834

00:20:43.080 --> 00:20:44.600 at high risk to become

NOTE Confidence: 0.9991834

00:20:44.600 --> 00:20:46.460 fluid overloaded very quickly.

NOTE Confidence: 0.9788536

00:20:46.919 --> 00:20:48.200 This means you must monitor

NOTE Confidence: 0.9788536

00:20:48.200 --> 00:20:49.960 the fluid responsiveness and fluid

NOTE Confidence: 0.9788536

00:20:49.960 --> 00:20:51.559 overload frequently and risk of

NOTE Confidence: 0.9788536

00:20:51.559 --> 00:20:53.900 fluid overload frequently. Also consider

NOTE Confidence: 0.9788536

00:20:53.960 --> 00:20:55.900 early use of vasopressor therapies

NOTE Confidence: 0.9788536

00:20:56.039 --> 00:20:57.500 as opposed to additional

NOTE Confidence: 0.99727345

00:20:57.960 --> 00:20:59.115 fluid boluses.

NOTE Confidence: 0.99979305

00:20:59.734 --> 00:21:02.135 Conservative fluid resuscitation is not

NOTE Confidence: 0.99979305

00:21:02.135 --> 00:21:02.635 unfamiliar

NOTE Confidence: 0.9929091

00:21:03.095 --> 00:21:04.934 to pediatric emergency medicine and

NOTE Confidence: 0.9929091

00:21:04.934 --> 00:21:06.075 critical care providers.

NOTE Confidence: 0.9994452

00:21:06.375 --> 00:21:07.654 Think about how we would  
NOTE Confidence: 0.9994452

00:21:07.654 --> 00:21:08.955 resuscitate a patient  
NOTE Confidence: 0.9996011

00:21:09.255 --> 00:21:10.234 in hypovolemic  
NOTE Confidence: 0.99837184

00:21:10.615 --> 00:21:12.154 shock as part of diabetic  
NOTE Confidence: 0.99837184

00:21:12.294 --> 00:21:12.794 ketoacidosis  
NOTE Confidence: 0.99912995

00:21:14.090 --> 00:21:15.850 To avoid or minimize the  
NOTE Confidence: 0.99912995

00:21:15.850 --> 00:21:17.950 risk of subsequent cerebral edema,  
NOTE Confidence: 0.99945647

00:21:18.330 --> 00:21:19.770 we would employ a similar  
NOTE Confidence: 0.99945647

00:21:19.770 --> 00:21:20.270 strategy  
NOTE Confidence: 0.9985063

00:21:20.730 --> 00:21:23.130 of smaller intravascular boluses with  
NOTE Confidence: 0.9985063

00:21:23.130 --> 00:21:24.350 more frequent reassessment  
NOTE Confidence: 0.96868205

00:21:24.890 --> 00:21:26.430 and early vasopressor  
NOTE Confidence: 0.9978737

00:21:26.890 --> 00:21:27.390 use.  
NOTE Confidence: 0.99699134

00:21:28.035 --> 00:21:29.715 Patients with systemic capillary leak  
NOTE Confidence: 0.99699134

00:21:29.715 --> 00:21:30.215 disease,  
NOTE Confidence: 0.9869881

00:21:30.675 --> 00:21:31.795 can be can have their

NOTE Confidence: 0.9869881

00:21:31.795 --> 00:21:33.395 cardiac output monitored the same

NOTE Confidence: 0.9869881

00:21:33.395 --> 00:21:34.994 way you would for other

NOTE Confidence: 0.9869881

00:21:34.994 --> 00:21:36.994 etiologies of shock, and that

NOTE Confidence: 0.9869881

00:21:36.994 --> 00:21:39.075 includes lactate, mixed venous oxygen

NOTE Confidence: 0.9869881

00:21:39.075 --> 00:21:39.575 saturation,

NOTE Confidence: 0.9457369

00:21:40.060 --> 00:21:41.540 urine output mental status, or

NOTE Confidence: 0.9457369

00:21:41.540 --> 00:21:43.119 if available at your institution

NOTE Confidence: 0.9457369

00:21:43.260 --> 00:21:43.760 echocardiography.

NOTE Confidence: 0.9544549

00:21:45.500 --> 00:21:47.280 Patients with s with SCLS

NOTE Confidence: 0.994627

00:21:47.580 --> 00:21:49.340 are at higher risk for

NOTE Confidence: 0.994627

00:21:49.340 --> 00:21:51.359 complications of resuscitation, however,

NOTE Confidence: 0.99839526

00:21:51.660 --> 00:21:53.600 and this can include compartment

NOTE Confidence: 0.99839526

00:21:53.660 --> 00:21:54.160 syndrome.

NOTE Confidence: 0.9996651

00:21:54.685 --> 00:21:56.145 Aggressive fluid resuscitation

NOTE Confidence: 0.98089075

00:21:56.845 --> 00:21:58.705 will lead to excessive extravascular

NOTE Confidence: 0.9996943

00:21:59.085 --> 00:22:00.145 leak of that fluid  
NOTE Confidence: 0.99924374

00:22:00.525 --> 00:22:01.825 into the extremities  
NOTE Confidence: 0.9815029

00:22:02.445 --> 00:22:04.445 and may quite quickly result  
NOTE Confidence: 0.9815029

00:22:04.445 --> 00:22:06.705 in loss of systemic pulses,  
NOTE Confidence: 0.9739204

00:22:07.410 --> 00:22:09.030 profound extremity pain,  
NOTE Confidence: 0.9489558

00:22:09.410 --> 00:22:09.730 and,  
NOTE Confidence: 0.9828901

00:22:10.290 --> 00:22:12.470 resulting all from compartment syndrome,  
NOTE Confidence: 0.9828901

00:22:12.770 --> 00:22:14.130 which needs to be addressed  
NOTE Confidence: 0.9828901

00:22:14.130 --> 00:22:14.630 immediately  
NOTE Confidence: 0.99922484

00:22:15.170 --> 00:22:17.330 by your orthopedic or plastic  
NOTE Confidence: 0.99922484

00:22:17.330 --> 00:22:17.830 surgery  
NOTE Confidence: 0.9994207

00:22:18.210 --> 00:22:18.710 services.  
NOTE Confidence: 0.9979789

00:22:19.090 --> 00:22:20.210 Patients are also at high  
NOTE Confidence: 0.9979789

00:22:20.210 --> 00:22:21.270 risk for other  
NOTE Confidence: 0.92837816

00:22:21.845 --> 00:22:24.505 vascular leak complications including abdominal  
NOTE Confidence: 0.92837816

00:22:24.645 --> 00:22:25.625 compartment syndrome,

NOTE Confidence: 0.8754031  
00:22:26.885 --> 00:22:27.385 pericardial  
NOTE Confidence: 0.90553904  
00:22:27.685 --> 00:22:28.185 effusions,  
NOTE Confidence: 0.9167549  
00:22:29.205 --> 00:22:30.505 or pleural effusions,  
NOTE Confidence: 0.99353397  
00:22:30.885 --> 00:22:32.565 which may all result in  
NOTE Confidence: 0.99353397  
00:22:32.565 --> 00:22:34.185 worse end organ dysfunction.  
NOTE Confidence: 0.96932787  
00:22:35.029 --> 00:22:36.230 These patients are also at  
NOTE Confidence: 0.96932787  
00:22:36.230 --> 00:22:38.090 high risk for coagulation abnormalities  
NOTE Confidence: 0.96932787  
00:22:38.149 --> 00:22:39.450 and deep venous thrombosis  
NOTE Confidence: 0.99326456  
00:22:39.909 --> 00:22:41.690 due to stasis of blood,  
NOTE Confidence: 0.9959818  
00:22:42.149 --> 00:22:44.309 and may require early treatment  
NOTE Confidence: 0.9959818  
00:22:44.309 --> 00:22:45.049 with anticoagulation,  
NOTE Confidence: 0.9932486  
00:22:45.669 --> 00:22:46.630 but that should be done  
NOTE Confidence: 0.9932486  
00:22:46.630 --> 00:22:48.570 in conjunction with your hospital  
NOTE Confidence: 0.9932486  
00:22:48.630 --> 00:22:49.130 policies.  
NOTE Confidence: 0.990325  
00:22:50.070 --> 00:22:51.795 The sharp drop in intravascular  
NOTE Confidence: 0.990325

00:22:52.015 --> 00:22:53.615 volume may also result in  
NOTE Confidence: 0.990325

00:22:53.615 --> 00:22:55.795 more profound acute kidney injury,  
NOTE Confidence: 0.9942751

00:22:56.175 --> 00:22:57.795 than typically seen in distributive  
NOTE Confidence: 0.9942751

00:22:57.855 --> 00:22:59.635 shock. All of these complications,  
NOTE Confidence: 0.99788636

00:23:00.335 --> 00:23:01.875 need to be monitored frequently  
NOTE Confidence: 0.99788636

00:23:01.935 --> 00:23:03.135 in the patient in the  
NOTE Confidence: 0.99788636

00:23:03.135 --> 00:23:04.035 leak phase.  
NOTE Confidence: 0.9996188

00:23:05.570 --> 00:23:07.430 Unfortunately, there are no specific  
NOTE Confidence: 0.9996188

00:23:07.490 --> 00:23:09.030 therapies for the leak phase.  
NOTE Confidence: 0.9954049

00:23:09.570 --> 00:23:10.850 There have been many tried,  
NOTE Confidence: 0.9954049

00:23:11.090 --> 00:23:12.850 but none are effective. Those  
NOTE Confidence: 0.9954049

00:23:12.850 --> 00:23:14.310 that have been tried include  
NOTE Confidence: 0.9954049

00:23:14.369 --> 00:23:16.150 IVIG, plasma exchange,  
NOTE Confidence: 0.9477649

00:23:16.645 --> 00:23:19.145 aminophylline or tributylamine, methylene blue,  
NOTE Confidence: 0.9477649

00:23:19.445 --> 00:23:20.424 targeted immunotherapies,  
NOTE Confidence: 0.9968346

00:23:21.205 --> 00:23:22.265 or even extracorporeal

NOTE Confidence: 0.9328362

00:23:22.645 --> 00:23:24.665 membranous oxygenation or ECMO.

NOTE Confidence: 0.9974457

00:23:25.125 --> 00:23:26.645 These therapies have not shown

NOTE Confidence: 0.9974457

00:23:26.645 --> 00:23:28.105 to be effective at reducing

NOTE Confidence: 0.9974457

00:23:28.244 --> 00:23:29.765 the intensity or duration of

NOTE Confidence: 0.9974457

00:23:29.765 --> 00:23:31.409 the leak phase and should

NOTE Confidence: 0.9974457

00:23:31.409 --> 00:23:33.090 only be leveraged for your

NOTE Confidence: 0.9974457

00:23:33.090 --> 00:23:34.850 patients in collaboration with local

NOTE Confidence: 0.9974457

00:23:34.850 --> 00:23:35.350 experts.

NOTE Confidence: 0.9875054

00:23:35.809 --> 00:23:37.250 The leak phase may last

NOTE Confidence: 0.9875054

00:23:37.250 --> 00:23:39.330 as little as several hours

NOTE Confidence: 0.9875054

00:23:39.330 --> 00:23:41.190 or up to seven days.

NOTE Confidence: 0.9875054

00:23:41.409 --> 00:23:43.350 They instantly stopped fluids.

NOTE Confidence: 0.9824386

00:23:43.825 --> 00:23:44.865 So they did not know

NOTE Confidence: 0.9824386

00:23:44.865 --> 00:23:46.465 what was going on. Their

NOTE Confidence: 0.9824386

00:23:46.465 --> 00:23:46.965 initial,

NOTE Confidence: 0.97108674

00:23:49.425 --> 00:23:51.185 assumption as almost all of  
NOTE Confidence: 0.97108674

00:23:51.185 --> 00:23:51.984 the kids that I know  
NOTE Confidence: 0.97108674

00:23:51.984 --> 00:23:53.984 with SCLS was nephrotic syndrome.  
NOTE Confidence: 0.97108674

00:23:53.984 --> 00:23:54.945 We that was the first  
NOTE Confidence: 0.97108674

00:23:54.945 --> 00:23:56.225 people we had in. We  
NOTE Confidence: 0.97108674

00:23:56.225 --> 00:23:57.505 actually were admitted on a  
NOTE Confidence: 0.97108674

00:23:57.505 --> 00:23:58.385 Sunday in the middle of  
NOTE Confidence: 0.97108674

00:23:58.385 --> 00:24:00.149 the night. Monday morning rounds,  
NOTE Confidence: 0.97108674

00:24:00.149 --> 00:24:01.210 the entire children's  
NOTE Confidence: 0.9996464

00:24:01.830 --> 00:24:03.429 nephrology team is in with  
NOTE Confidence: 0.9996464

00:24:03.429 --> 00:24:03.929 us  
NOTE Confidence: 0.9629996

00:24:04.309 --> 00:24:05.909 taking all of that. I  
NOTE Confidence: 0.9629996

00:24:05.909 --> 00:24:07.269 will say during some of  
NOTE Confidence: 0.9629996

00:24:07.269 --> 00:24:09.269 those previous leaks, her urine  
NOTE Confidence: 0.9629996

00:24:09.269 --> 00:24:10.950 was tested for protein. So  
NOTE Confidence: 0.9629996

00:24:10.950 --> 00:24:12.325 that was always top of

NOTE Confidence: 0.9629996  
00:24:12.325 --> 00:24:14.325 mind awareness was checking kidney  
NOTE Confidence: 0.9629996  
00:24:14.325 --> 00:24:15.684 functions, but it was always  
NOTE Confidence: 0.9629996  
00:24:15.684 --> 00:24:16.184 perfect.  
NOTE Confidence: 0.99473345  
00:24:17.044 --> 00:24:18.505 So we did do imaging,  
NOTE Confidence: 0.9786311  
00:24:19.365 --> 00:24:21.044 of her kidneys. Everything was  
NOTE Confidence: 0.9786311  
00:24:21.044 --> 00:24:22.744 perfect. No protein in urine.  
NOTE Confidence: 0.9786311  
00:24:23.044 --> 00:24:24.480 And at this time, she  
NOTE Confidence: 0.9786311  
00:24:24.480 --> 00:24:26.400 started dumping that IV fluid,  
NOTE Confidence: 0.9786311  
00:24:26.400 --> 00:24:27.600 and she gained ten percent  
NOTE Confidence: 0.9786311  
00:24:27.600 --> 00:24:28.740 of her body weight  
NOTE Confidence: 0.99941474  
00:24:29.040 --> 00:24:30.420 within about twelve hours.  
NOTE Confidence: 0.9994537  
00:24:31.040 --> 00:24:32.740 The recovery phase of  
NOTE Confidence: 0.96949303  
00:24:33.119 --> 00:24:34.960 systemic capillary leak disease may  
NOTE Confidence: 0.96949303  
00:24:34.960 --> 00:24:35.780 start suddenly.  
NOTE Confidence: 0.9997267  
00:24:36.240 --> 00:24:37.815 And this is associated with  
NOTE Confidence: 0.9997267

00:24:38.054 --> 00:24:38.875 rapid recruitment  
NOTE Confidence: 0.99884063

00:24:39.335 --> 00:24:40.615 of all of the volume  
NOTE Confidence: 0.99884063

00:24:40.615 --> 00:24:41.654 that had leaked out of  
NOTE Confidence: 0.99884063

00:24:41.654 --> 00:24:43.575 the patient's blood vessels back  
NOTE Confidence: 0.99884063

00:24:43.575 --> 00:24:44.554 into the intravascular  
NOTE Confidence: 0.9996258

00:24:44.855 --> 00:24:45.355 space.  
NOTE Confidence: 0.99402833

00:24:45.894 --> 00:24:47.674 This may be associated with  
NOTE Confidence: 0.99402833

00:24:47.894 --> 00:24:49.434 flash pulmonary edema  
NOTE Confidence: 0.9885055

00:24:49.734 --> 00:24:51.914 due to increased hydrostatic pressures  
NOTE Confidence: 0.99977666

00:24:52.549 --> 00:24:54.549 and heart failure due to  
NOTE Confidence: 0.99977666

00:24:54.549 --> 00:24:55.049 sudden  
NOTE Confidence: 0.9980724

00:24:55.350 --> 00:24:55.850 circulatory  
NOTE Confidence: 0.9993478

00:24:56.549 --> 00:24:57.049 overload.  
NOTE Confidence: 0.9951116

00:24:57.590 --> 00:24:58.790 This needs to be monitored  
NOTE Confidence: 0.9951116

00:24:58.790 --> 00:25:00.630 carefully in these patients and  
NOTE Confidence: 0.9951116

00:25:00.630 --> 00:25:02.090 treat treated with aggressive

NOTE Confidence: 0.9974105

00:25:02.549 --> 00:25:03.049 diuresis.

NOTE Confidence: 0.9476621

00:25:04.070 --> 00:25:05.690 Also, during hospital stays,

NOTE Confidence: 0.9970226

00:25:06.225 --> 00:25:08.385 patients should have consults for

NOTE Confidence: 0.9970226

00:25:08.385 --> 00:25:10.165 dedicated pediatric subspecialties.

NOTE Confidence: 0.99773264

00:25:10.705 --> 00:25:12.165 These include rheumatology

NOTE Confidence: 0.96696615

00:25:12.945 --> 00:25:13.845 and or

NOTE Confidence: 0.9642444

00:25:14.225 --> 00:25:15.445 allergy and immunology,

NOTE Confidence: 0.9810586

00:25:16.465 --> 00:25:18.165 as well as those consults

NOTE Confidence: 0.9810586

00:25:18.225 --> 00:25:19.925 dictated by end organ dysfunction,

NOTE Confidence: 0.99838585

00:25:20.570 --> 00:25:22.510 such as infectious disease, nephrology,

NOTE Confidence: 0.99838585

00:25:22.809 --> 00:25:23.309 cardiology,

NOTE Confidence: 0.9032239

00:25:24.730 --> 00:25:26.650 or or neurology or other,

NOTE Confidence: 0.9983073

00:25:27.050 --> 00:25:27.550 services.

NOTE Confidence: 0.9948967

00:25:28.250 --> 00:25:29.869 In conjunction with these services,

NOTE Confidence: 0.9988916

00:25:30.250 --> 00:25:31.390 tertiary evaluation

NOTE Confidence: 0.99634624

00:25:32.010 --> 00:25:33.950 for systemic capillary leak disease  
NOTE Confidence: 0.99634624

00:25:34.010 --> 00:25:34.510 include  
NOTE Confidence: 0.9142726

00:25:35.375 --> 00:25:35.875 immunoelectrophoresis  
NOTE Confidence: 0.94997114

00:25:36.654 --> 00:25:38.434 to look for the monoclonal  
NOTE Confidence: 0.94997114

00:25:38.654 --> 00:25:40.975 gammopathy of undetermined significance both  
NOTE Confidence: 0.94997114

00:25:40.975 --> 00:25:41.774 in the blood and the  
NOTE Confidence: 0.94997114

00:25:41.774 --> 00:25:42.274 urine,  
NOTE Confidence: 0.8730832

00:25:42.894 --> 00:25:43.955 urine analyses,  
NOTE Confidence: 0.97674406

00:25:44.894 --> 00:25:47.154 to, rule out nephrotic syndrome,  
NOTE Confidence: 0.9302926

00:25:47.670 --> 00:25:48.170 targeted,  
NOTE Confidence: 0.94046444

00:25:48.630 --> 00:25:50.330 tests to rule out anaphylaxis  
NOTE Confidence: 0.94046444

00:25:50.630 --> 00:25:52.550 such as negative tryptase or  
NOTE Confidence: 0.94046444

00:25:52.550 --> 00:25:54.170 rule out hereditary angioedema  
NOTE Confidence: 0.96779275

00:25:54.550 --> 00:25:56.250 such as c one esterase  
NOTE Confidence: 0.96779275

00:25:56.310 --> 00:25:58.470 inhibitor levels. If there's cardiac  
NOTE Confidence: 0.96779275

00:25:58.470 --> 00:25:58.970 dysfunction,

NOTE Confidence: 0.97388816

00:25:59.590 --> 00:26:01.450 you may consider advanced imaging

NOTE Confidence: 0.97388816

00:26:01.750 --> 00:26:02.970 of of heart function.

NOTE Confidence: 0.983278

00:26:03.455 --> 00:26:04.575 And if available at your

NOTE Confidence: 0.983278

00:26:04.575 --> 00:26:05.075 institution,

NOTE Confidence: 0.96986014

00:26:05.695 --> 00:26:07.375 rapid whole genome or other

NOTE Confidence: 0.96986014

00:26:07.375 --> 00:26:09.695 targeted genetic testing. She never

NOTE Confidence: 0.96986014

00:26:09.695 --> 00:26:10.675 went to PICU.

NOTE Confidence: 0.9803024

00:26:11.455 --> 00:26:12.494 She all she stayed on

NOTE Confidence: 0.9803024

00:26:12.494 --> 00:26:14.255 the main floor. Her albumin

NOTE Confidence: 0.9803024

00:26:14.255 --> 00:26:15.875 was always low. They never

NOTE Confidence: 0.9803024

00:26:15.935 --> 00:26:17.630 gave her replacement albumin. They

NOTE Confidence: 0.9803024

00:26:17.630 --> 00:26:18.750 waited to see if she

NOTE Confidence: 0.9803024

00:26:18.750 --> 00:26:20.269 could pee all of that

NOTE Confidence: 0.9803024

00:26:20.269 --> 00:26:21.630 extra fluid out, which she

NOTE Confidence: 0.9803024

00:26:21.630 --> 00:26:22.130 did.

NOTE Confidence: 0.99804074

00:26:22.750 --> 00:26:24.130 But Monday was nephrology.  
NOTE Confidence: 0.9989494

00:26:25.149 --> 00:26:27.090 Tuesday, we met with endocrinology  
NOTE Confidence: 0.93538713

00:26:27.470 --> 00:26:29.330 and explored Addison's disease,  
NOTE Confidence: 0.939252

00:26:29.914 --> 00:26:32.315 again, having some symptoms but  
NOTE Confidence: 0.939252

00:26:32.315 --> 00:26:33.294 missing others.  
NOTE Confidence: 0.97537184

00:26:34.315 --> 00:26:36.395 That cleared up. We also  
NOTE Confidence: 0.97537184

00:26:36.395 --> 00:26:39.054 saw allergy immunology for angioedema.  
NOTE Confidence: 0.9845594

00:26:39.994 --> 00:26:41.434 He quickly was like, this  
NOTE Confidence: 0.9845594

00:26:41.434 --> 00:26:42.575 is not angioedema,  
NOTE Confidence: 0.99507385

00:26:42.955 --> 00:26:44.075 but they still ruled it  
NOTE Confidence: 0.99507385

00:26:44.075 --> 00:26:44.460 out.  
NOTE Confidence: 0.999442

00:26:45.419 --> 00:26:46.399 And then the last  
NOTE Confidence: 0.97655976

00:26:46.940 --> 00:26:49.200 specialty that explored was gastro.  
NOTE Confidence: 0.97655976

00:26:49.260 --> 00:26:50.619 I have celiac, so we  
NOTE Confidence: 0.97655976

00:26:50.619 --> 00:26:52.640 thought maybe something's going on.  
NOTE Confidence: 0.97655976

00:26:52.940 --> 00:26:54.539 Maybe the protein is leaking

NOTE Confidence: 0.97655976  
00:26:54.539 --> 00:26:56.140 through the GI system. So  
NOTE Confidence: 0.97655976  
00:26:56.140 --> 00:26:57.500 she had stool samples sent  
NOTE Confidence: 0.97655976  
00:26:57.500 --> 00:26:59.085 off, all of that, and  
NOTE Confidence: 0.97655976  
00:26:59.085 --> 00:27:00.684 that came back clear. And  
NOTE Confidence: 0.97655976  
00:27:00.684 --> 00:27:02.044 it was not until the  
NOTE Confidence: 0.97655976  
00:27:02.044 --> 00:27:02.544 rheumatology  
NOTE Confidence: 0.95336026  
00:27:02.924 --> 00:27:04.065 team came in,  
NOTE Confidence: 0.99455976  
00:27:04.765 --> 00:27:05.804 and it was actually the  
NOTE Confidence: 0.99455976  
00:27:05.804 --> 00:27:06.304 fellow  
NOTE Confidence: 0.70308614  
00:27:06.845 --> 00:27:07.345 who,  
NOTE Confidence: 0.958925  
00:27:09.005 --> 00:27:10.605 took the assessment and presented  
NOTE Confidence: 0.958925  
00:27:10.605 --> 00:27:11.965 it to her attending. And  
NOTE Confidence: 0.958925  
00:27:11.965 --> 00:27:13.244 it was the attending, doctor  
NOTE Confidence: 0.958925  
00:27:13.244 --> 00:27:14.304 Rebecca Sedoun,  
NOTE Confidence: 0.9660792  
00:27:14.950 --> 00:27:16.710 who came in maybe five  
NOTE Confidence: 0.9660792

00:27:16.710 --> 00:27:18.230 minutes later. Yeah. And she

NOTE Confidence: 0.9660792

00:27:18.230 --> 00:27:19.430 was like, your daughter is

NOTE Confidence: 0.9660792

00:27:19.430 --> 00:27:20.170 a unicorn.

NOTE Confidence: 0.9811997

00:27:21.110 --> 00:27:22.470 I have never seen this.

NOTE Confidence: 0.9811997

00:27:22.470 --> 00:27:23.910 I will probably never see

NOTE Confidence: 0.9811997

00:27:23.910 --> 00:27:24.650 this again.

NOTE Confidence: 0.9996004

00:27:25.270 --> 00:27:27.110 And she told us the

NOTE Confidence: 0.9996004

00:27:27.110 --> 00:27:27.610 diagnosis.

NOTE Confidence: 0.99789965

00:27:28.985 --> 00:27:30.985 This table summarizes the inpatient

NOTE Confidence: 0.99789965

00:27:30.985 --> 00:27:33.405 treatment differences between the differential

NOTE Confidence: 0.99789965

00:27:33.465 --> 00:27:33.965 diagnosis.

NOTE Confidence: 0.99405557

00:27:34.585 --> 00:27:35.945 Again, I would note the

NOTE Confidence: 0.99405557

00:27:35.945 --> 00:27:38.585 conservative fluid resuscitation and early

NOTE Confidence: 0.99405557

00:27:38.585 --> 00:27:39.885 vasopressor therapy

NOTE Confidence: 0.99826276

00:27:40.185 --> 00:27:41.865 for patients with systemic capillary

NOTE Confidence: 0.99826276

00:27:41.865 --> 00:27:43.485 leak disease to help minimize

NOTE Confidence: 0.9998715

00:27:44.210 --> 00:27:44.869 the complications

NOTE Confidence: 0.99972606

00:27:45.570 --> 00:27:47.670 of the recovery phase.

NOTE Confidence: 0.99737597

00:27:48.130 --> 00:27:49.270 Although there are no

NOTE Confidence: 0.9998081

00:27:49.650 --> 00:27:51.510 effective therapies to decrease

NOTE Confidence: 0.99961793

00:27:51.810 --> 00:27:53.250 the duration or intensity of

NOTE Confidence: 0.99961793

00:27:53.250 --> 00:27:54.609 the leak phase, there are

NOTE Confidence: 0.99961793

00:27:54.609 --> 00:27:56.150 effective therapies to

NOTE Confidence: 0.98360085

00:27:56.625 --> 00:27:58.305 reduce the risk of having

NOTE Confidence: 0.98360085

00:27:58.305 --> 00:28:00.385 another leak phase. In addition

NOTE Confidence: 0.98360085

00:28:00.385 --> 00:28:02.065 to all of the pediatric

NOTE Confidence: 0.98360085

00:28:02.065 --> 00:28:04.145 subspecialty follow-up based on organ

NOTE Confidence: 0.98360085

00:28:04.145 --> 00:28:05.744 injury, children should be followed

NOTE Confidence: 0.98360085

00:28:05.744 --> 00:28:06.405 by rheumatology

NOTE Confidence: 0.976728

00:28:06.785 --> 00:28:08.085 or allergy immunology

NOTE Confidence: 0.9984091

00:28:08.720 --> 00:28:10.180 depending on who's most comfortable

NOTE Confidence: 0.9951509

00:28:10.560 --> 00:28:11.060 prescribing  
NOTE Confidence: 0.9313074

00:28:11.440 --> 00:28:12.660 intravenous immunoglobulin  
NOTE Confidence: 0.89982504

00:28:13.119 --> 00:28:13.940 or IVIG,  
NOTE Confidence: 0.9997255

00:28:14.880 --> 00:28:15.859 to these patients.  
NOTE Confidence: 0.99966294

00:28:16.160 --> 00:28:17.840 IVIG is the first line  
NOTE Confidence: 0.99966294

00:28:17.840 --> 00:28:18.340 treatment  
NOTE Confidence: 0.99964595

00:28:18.640 --> 00:28:19.619 for preventing  
NOTE Confidence: 0.9992309

00:28:20.080 --> 00:28:21.920 leak episodes in patients with  
NOTE Confidence: 0.9992309

00:28:21.920 --> 00:28:22.420 SCLS.  
NOTE Confidence: 0.89684486

00:28:23.225 --> 00:28:24.125 It significantly,  
NOTE Confidence: 0.9857612

00:28:25.145 --> 00:28:27.165 reduces relapse rates and increases  
NOTE Confidence: 0.9857612

00:28:27.225 --> 00:28:28.205 ten year survival  
NOTE Confidence: 0.99016255

00:28:28.744 --> 00:28:30.285 up to one hundred percent,  
NOTE Confidence: 0.99987906

00:28:30.665 --> 00:28:31.405 in adults.  
NOTE Confidence: 0.96287155

00:28:32.585 --> 00:28:34.825 Typically, IVIG doses are starting  
NOTE Confidence: 0.96287155

00:28:34.825 --> 00:28:35.940 on the higher end,

NOTE Confidence: 0.97595215

00:28:36.419 --> 00:28:38.039 of one to two kilograms

NOTE Confidence: 0.97595215

00:28:38.100 --> 00:28:40.120 per month and then titrated

NOTE Confidence: 0.97595215

00:28:40.259 --> 00:28:42.360 in close collaboration with rheumatology

NOTE Confidence: 0.97595215

00:28:42.659 --> 00:28:44.519 or allergy immunology experts

NOTE Confidence: 0.95879287

00:28:44.980 --> 00:28:46.360 to the, tolerated,

NOTE Confidence: 0.99975127

00:28:46.820 --> 00:28:47.320 dose.

NOTE Confidence: 0.991066

00:28:47.860 --> 00:28:49.255 If patients are unable to

NOTE Confidence: 0.991066

00:28:49.255 --> 00:28:51.255 tolerate IVIG, and that may

NOTE Confidence: 0.991066

00:28:51.255 --> 00:28:53.015 be due to headaches or

NOTE Confidence: 0.991066

00:28:53.015 --> 00:28:54.875 migraines associated with infusion,

NOTE Confidence: 0.977104

00:28:55.335 --> 00:28:57.255 other infusion reactions, or the

NOTE Confidence: 0.977104

00:28:57.255 --> 00:28:58.955 cost or the time required,

NOTE Confidence: 0.99354166

00:28:59.575 --> 00:29:00.855 to administer this drug at

NOTE Confidence: 0.99354166

00:29:00.855 --> 00:29:02.940 infusion centers, they may benefit

NOTE Confidence: 0.99354166

00:29:03.000 --> 00:29:03.820 from subcutaneous

NOTE Confidence: 0.99960494

00:29:04.280 --> 00:29:04.780 immunoglobulin  
NOTE Confidence: 0.868418

00:29:05.240 --> 00:29:05.980 or SCIG  
NOTE Confidence: 0.9892539

00:29:06.600 --> 00:29:07.560 at a dose of eighty  
NOTE Confidence: 0.9892539

00:29:07.560 --> 00:29:09.240 milligrams per kilogram up to  
NOTE Confidence: 0.9892539

00:29:09.240 --> 00:29:10.300 three times a week.  
NOTE Confidence: 0.9636345

00:29:11.240 --> 00:29:12.780 Other older therapies,  
NOTE Confidence: 0.98116744

00:29:13.160 --> 00:29:15.740 for SCLS include terbutaline or  
NOTE Confidence: 0.8989967

00:29:16.054 --> 00:29:16.554 theophylline.  
NOTE Confidence: 0.90083647

00:29:17.255 --> 00:29:17.755 In,  
NOTE Confidence: 0.9840908

00:29:18.535 --> 00:29:20.934 other therapies for SCLS include  
NOTE Confidence: 0.9840908

00:29:20.934 --> 00:29:21.434 terbutaline  
NOTE Confidence: 0.9906523

00:29:21.735 --> 00:29:22.475 or theophylline.  
NOTE Confidence: 0.9979032

00:29:22.934 --> 00:29:24.294 These have been used together  
NOTE Confidence: 0.9979032

00:29:24.294 --> 00:29:25.035 or separately,  
NOTE Confidence: 0.9700526

00:29:25.575 --> 00:29:27.015 and are really much, much  
NOTE Confidence: 0.9700526

00:29:27.015 --> 00:29:28.554 less effective than IVIG.

NOTE Confidence: 0.9619124  
00:29:29.580 --> 00:29:30.320 Failed therapies,  
NOTE Confidence: 0.86086756  
00:29:30.700 --> 00:29:32.059 are listed here but includes  
NOTE Confidence: 0.86086756  
00:29:32.059 --> 00:29:33.279 steroids, diuretics,  
NOTE Confidence: 0.9847644  
00:29:34.299 --> 00:29:34.799 leukotriene  
NOTE Confidence: 0.96836364  
00:29:35.100 --> 00:29:37.419 modifying agents, and other drugs  
NOTE Confidence: 0.96836364  
00:29:37.419 --> 00:29:39.100 or herbal supplements are not  
NOTE Confidence: 0.96836364  
00:29:39.100 --> 00:29:40.700 recommended and they do not  
NOTE Confidence: 0.96836364  
00:29:40.700 --> 00:29:41.200 prevent  
NOTE Confidence: 0.99932784  
00:29:41.674 --> 00:29:43.215 episodes of leak.  
NOTE Confidence: 0.9978513  
00:29:44.315 --> 00:29:45.595 So here is our completed  
NOTE Confidence: 0.9978513  
00:29:45.595 --> 00:29:46.095 table,  
NOTE Confidence: 0.9915834  
00:29:46.475 --> 00:29:48.395 which highlights the differences between  
NOTE Confidence: 0.9915834  
00:29:48.395 --> 00:29:50.174 systemic capillary leak disease  
NOTE Confidence: 0.960809  
00:29:50.635 --> 00:29:52.335 and, other diagnostic  
NOTE Confidence: 0.99957865  
00:29:52.635 --> 00:29:53.135 considerations  
NOTE Confidence: 0.97548836

00:29:53.515 --> 00:29:55.440 in the short term, long  
NOTE Confidence: 0.97548836

00:29:55.440 --> 00:29:57.140 term, and response to treatment?  
NOTE Confidence: 0.9744647

00:29:58.320 --> 00:30:00.260 At almost every single pediatric  
NOTE Confidence: 0.9744647

00:30:00.320 --> 00:30:01.920 case, it's like reading her  
NOTE Confidence: 0.9744647

00:30:01.920 --> 00:30:02.900 MyChart notes.  
NOTE Confidence: 0.8962443

00:30:03.760 --> 00:30:05.520 Child presents to the ED  
NOTE Confidence: 0.8962443

00:30:05.520 --> 00:30:07.840 with court with, cold like  
NOTE Confidence: 0.8962443

00:30:07.840 --> 00:30:08.340 symptoms,  
NOTE Confidence: 0.985326

00:30:09.165 --> 00:30:10.765 high heart rate, low blood  
NOTE Confidence: 0.985326

00:30:10.765 --> 00:30:12.365 pressure, low albumin. And so  
NOTE Confidence: 0.985326

00:30:12.365 --> 00:30:13.485 to me, that was so  
NOTE Confidence: 0.985326

00:30:13.485 --> 00:30:15.405 frustrating as a parent that  
NOTE Confidence: 0.985326

00:30:15.405 --> 00:30:16.525 how could it be these  
NOTE Confidence: 0.985326

00:30:16.525 --> 00:30:17.425 three or four  
NOTE Confidence: 0.9780644

00:30:18.045 --> 00:30:18.545 identical  
NOTE Confidence: 0.85326797

00:30:18.925 --> 00:30:20.705 simple clinical presentations

NOTE Confidence: 0.8942493

00:30:21.565 --> 00:30:22.765 and no one has heard

NOTE Confidence: 0.8942493

00:30:22.765 --> 00:30:23.425 of this.

NOTE Confidence: 0.9632832

00:30:24.720 --> 00:30:26.160 And that's where my passion

NOTE Confidence: 0.9632832

00:30:26.160 --> 00:30:28.340 comes from with raising awareness.

NOTE Confidence: 0.99722755

00:30:28.800 --> 00:30:30.580 So those first three episodes,

NOTE Confidence: 0.9903728

00:30:31.360 --> 00:30:33.140 I think I was doing

NOTE Confidence: 0.981773

00:30:33.680 --> 00:30:35.280 I reacted how any mom

NOTE Confidence: 0.981773

00:30:35.280 --> 00:30:36.320 would. I was still very

NOTE Confidence: 0.981773

00:30:36.320 --> 00:30:37.360 concerned, and I was still

NOTE Confidence: 0.981773

00:30:37.360 --> 00:30:38.755 calling the nurse line. I

NOTE Confidence: 0.981773

00:30:38.755 --> 00:30:39.875 feel like I did everything

NOTE Confidence: 0.981773

00:30:39.875 --> 00:30:40.375 right.

NOTE Confidence: 0.9840323

00:30:41.155 --> 00:30:42.195 If I were to whisper

NOTE Confidence: 0.9840323

00:30:42.195 --> 00:30:44.115 in my ear back with

NOTE Confidence: 0.9840323

00:30:44.115 --> 00:30:46.034 the influenza a, that really

NOTE Confidence: 0.9840323

00:30:46.034 --> 00:30:46.534 bad  
NOTE Confidence: 0.90370744

00:30:46.835 --> 00:30:48.135 third fourth week,  
NOTE Confidence: 0.9171861

00:30:48.515 --> 00:30:50.275 I would have insisted on  
NOTE Confidence: 0.9171861

00:30:50.275 --> 00:30:51.900 blood work. And but it's  
NOTE Confidence: 0.9171861

00:30:51.900 --> 00:30:53.920 really hard to convince pediatric  
NOTE Confidence: 0.9171861

00:30:54.060 --> 00:30:54.560 providers  
NOTE Confidence: 0.9822388

00:30:54.860 --> 00:30:56.300 to do labs. And it's  
NOTE Confidence: 0.9822388

00:30:56.300 --> 00:30:57.500 also hard because I don't  
NOTE Confidence: 0.9822388

00:30:57.660 --> 00:30:58.700 you don't wanna do labs  
NOTE Confidence: 0.9822388

00:30:58.700 --> 00:30:59.500 on your kids. It's a  
NOTE Confidence: 0.9822388

00:30:59.500 --> 00:31:00.940 nightmare. Right? It's terrible. They're  
NOTE Confidence: 0.9822388

00:31:00.940 --> 00:31:02.460 screaming and crying. So it's  
NOTE Confidence: 0.9822388

00:31:02.460 --> 00:31:04.060 not like you necessarily wanna  
NOTE Confidence: 0.9822388

00:31:04.060 --> 00:31:05.245 ask for blood work. And  
NOTE Confidence: 0.9822388

00:31:05.245 --> 00:31:06.925 when your pediatrician who you  
NOTE Confidence: 0.9822388

00:31:06.925 --> 00:31:08.465 trust and knows your child

NOTE Confidence: 0.95182675  
00:31:08.925 --> 00:31:10.465 isn't saying you need it,  
NOTE Confidence: 0.99027705  
00:31:10.925 --> 00:31:12.285 you know, but we both  
NOTE Confidence: 0.99027705  
00:31:12.285 --> 00:31:13.645 knew something was going on.  
NOTE Confidence: 0.99027705  
00:31:13.645 --> 00:31:15.565 The frustration was there probably  
NOTE Confidence: 0.99027705  
00:31:15.565 --> 00:31:16.845 since the second week. I  
NOTE Confidence: 0.99027705  
00:31:16.845 --> 00:31:17.585 was like,  
NOTE Confidence: 0.9498404  
00:31:17.910 --> 00:31:19.590 this is not just unknown  
NOTE Confidence: 0.9498404  
00:31:19.590 --> 00:31:21.270 eyes falling, this random eyes  
NOTE Confidence: 0.9498404  
00:31:21.270 --> 00:31:22.470 falling. Like, there has to  
NOTE Confidence: 0.9498404  
00:31:22.470 --> 00:31:23.210 be a reason.  
NOTE Confidence: 0.90414864  
00:31:23.670 --> 00:31:25.050 Education is huge.  
NOTE Confidence: 0.9595378  
00:31:25.510 --> 00:31:26.790 And and bring it to  
NOTE Confidence: 0.9595378  
00:31:26.790 --> 00:31:28.390 your doctors because a lot  
NOTE Confidence: 0.9595378  
00:31:28.390 --> 00:31:29.750 of them, you have to  
NOTE Confidence: 0.9595378  
00:31:29.750 --> 00:31:32.345 become, like, your own expert  
NOTE Confidence: 0.9595378

00:31:32.345 --> 00:31:34.024 on it. They're not gonna  
NOTE Confidence: 0.9595378

00:31:34.024 --> 00:31:35.385 have read as many papers  
NOTE Confidence: 0.9595378

00:31:35.385 --> 00:31:36.264 as you on it most  
NOTE Confidence: 0.9595378

00:31:36.264 --> 00:31:36.764 likely.  
NOTE Confidence: 0.9846986

00:31:37.385 --> 00:31:38.745 They're you know, they may  
NOTE Confidence: 0.9846986

00:31:38.745 --> 00:31:40.365 not know who to contact,  
NOTE Confidence: 0.9846986

00:31:40.585 --> 00:31:42.605 but it's a very small  
NOTE Confidence: 0.9846986

00:31:42.664 --> 00:31:43.164 circle  
NOTE Confidence: 0.9998933

00:31:43.660 --> 00:31:44.400 with this  
NOTE Confidence: 0.96506846

00:31:44.780 --> 00:31:46.540 disease. And we feel very,  
NOTE Confidence: 0.96506846

00:31:46.540 --> 00:31:48.480 very blessed for doctor Drury's  
NOTE Confidence: 0.96506846

00:31:48.540 --> 00:31:49.040 research,  
NOTE Confidence: 0.949506

00:31:49.500 --> 00:31:50.540 which I know is more  
NOTE Confidence: 0.949506

00:31:50.540 --> 00:31:51.980 adult based and even more  
NOTE Confidence: 0.949506

00:31:51.980 --> 00:31:54.080 blessed that, doctor Pierce, you're  
NOTE Confidence: 0.9791402

00:31:54.380 --> 00:31:55.580 kinda taking the torch and

NOTE Confidence: 0.9791402  
00:31:55.580 --> 00:31:56.780 and moving the needle, if  
NOTE Confidence: 0.9791402  
00:31:56.780 --> 00:31:57.820 you will, here in the  
NOTE Confidence: 0.9791402  
00:31:57.820 --> 00:31:59.674 United States for SCLS.  
NOTE Confidence: 0.9786385  
00:32:00.695 --> 00:32:01.975 So, yeah, I think just  
NOTE Confidence: 0.9786385  
00:32:01.975 --> 00:32:04.235 advocate and educate and read  
NOTE Confidence: 0.9298807  
00:32:05.414 --> 00:32:06.774 and talk to one another  
NOTE Confidence: 0.9298807  
00:32:06.774 --> 00:32:07.514 as parents.  
NOTE Confidence: 0.99398476  
00:32:08.534 --> 00:32:09.495 So I hope that I  
NOTE Confidence: 0.99398476  
00:32:09.495 --> 00:32:11.355 have convinced you to consider  
NOTE Confidence: 0.99398476  
00:32:11.414 --> 00:32:13.195 systemic capillary leak disease,  
NOTE Confidence: 0.99957734  
00:32:13.660 --> 00:32:15.260 in children that have one  
NOTE Confidence: 0.99957734  
00:32:15.260 --> 00:32:17.900 or more unexplained episodes of  
NOTE Confidence: 0.99957734  
00:32:17.900 --> 00:32:19.200 hypovolemic shock.  
NOTE Confidence: 0.9864593  
00:32:19.500 --> 00:32:21.260 This is especially important for  
NOTE Confidence: 0.9864593  
00:32:21.260 --> 00:32:22.320 those that have,  
NOTE Confidence: 0.9998201

00:32:22.780 --> 00:32:23.760 episodes of  
NOTE Confidence: 0.97750306

00:32:24.060 --> 00:32:26.025 culture negative septic shock,  
NOTE Confidence: 0.99020547

00:32:26.585 --> 00:32:28.745 to be considering systemic capillary  
NOTE Confidence: 0.99020547

00:32:28.745 --> 00:32:29.485 leak disease.  
NOTE Confidence: 0.99109316

00:32:29.785 --> 00:32:31.145 And that's because many children  
NOTE Confidence: 0.99109316

00:32:31.145 --> 00:32:32.185 will have more than one  
NOTE Confidence: 0.99109316

00:32:32.185 --> 00:32:33.465 episode of leak before this  
NOTE Confidence: 0.99109316

00:32:33.465 --> 00:32:34.125 is diagnosed,  
NOTE Confidence: 0.99975616

00:32:34.745 --> 00:32:35.725 and that's preventable.  
NOTE Confidence: 0.9515804

00:32:36.345 --> 00:32:37.465 I hopefully, I've convinced you  
NOTE Confidence: 0.9515804

00:32:37.465 --> 00:32:38.525 that there's effective  
NOTE Confidence: 0.9987575

00:32:38.905 --> 00:32:41.005 outpatient treatment that can reduce  
NOTE Confidence: 0.99074465

00:32:41.305 --> 00:32:42.970 the episodes of leak or  
NOTE Confidence: 0.99074465

00:32:42.970 --> 00:32:44.490 prevent the episodes of leak  
NOTE Confidence: 0.99074465

00:32:44.490 --> 00:32:46.350 with IVIG or SCIG.  
NOTE Confidence: 0.9545798

00:32:47.610 --> 00:32:48.830 An increased awareness,

NOTE Confidence: 0.99383605  
00:32:49.370 --> 00:32:51.610 of pediatric emergency medicine and  
NOTE Confidence: 0.99383605  
00:32:51.610 --> 00:32:52.750 critical care providers  
NOTE Confidence: 0.9989128  
00:32:53.370 --> 00:32:54.730 may decrease the number of  
NOTE Confidence: 0.9989128  
00:32:54.730 --> 00:32:56.570 episodes of leak before there's  
NOTE Confidence: 0.9989128  
00:32:56.570 --> 00:32:57.230 a diagnosis.  
NOTE Confidence: 0.95363635  
00:32:58.385 --> 00:32:59.505 I continue to run a  
NOTE Confidence: 0.95363635  
00:32:59.505 --> 00:33:00.945 lab focused on blood vessel  
NOTE Confidence: 0.95363635  
00:33:00.945 --> 00:33:02.465 dysfunction in critically ill children  
NOTE Confidence: 0.95363635  
00:33:02.465 --> 00:33:03.985 and am actively invested and  
NOTE Confidence: 0.95363635  
00:33:03.985 --> 00:33:04.485 involved  
NOTE Confidence: 0.9898478  
00:33:04.865 --> 00:33:06.885 in children diagnosed with capillary  
NOTE Confidence: 0.9898478  
00:33:06.945 --> 00:33:07.684 leak syndrome.  
NOTE Confidence: 0.991279  
00:33:07.985 --> 00:33:09.105 I'd be happy to hear  
NOTE Confidence: 0.991279  
00:33:09.105 --> 00:33:10.865 from providers or patients at  
NOTE Confidence: 0.991279  
00:33:10.865 --> 00:33:12.725 the email address listed there.