

1

00:00:03,560 --> 00:00:06,000

Great Ormond Street Children's Hospital has helped develop cures

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00:00:06,000 --> 00:00:09,880

for many conditions which just a few years ago were untreatable.

3

00:00:09,880 --> 00:00:13,480

But for medical science to move forward,

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00:00:13,480 --> 00:00:15,720

doctors must be able to experiment.

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00:00:15,720 --> 00:00:20,560

We've done a very small number of patients with this condition in England.

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00:00:20,560 --> 00:00:25,400

If you sign the consent form, you're basically signing a contract with uncertainty.

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00:00:25,400 --> 00:00:31,080

Experimental surgery on children raises difficult ethical dilemmas.

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00:00:31,080 --> 00:00:34,280

Whenever you have any patient, it's trying to make the right decision

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00:00:34,280 --> 00:00:37,360

and when they are so severe,

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00:00:37,360 --> 00:00:41,400

possibly the right decision is not to operate on them.

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00:00:41,400 --> 00:00:44,240

For children with conditions which have no known cure,

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00:00:44,240 --> 00:00:47,440

these experiments are their only hope.

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00:00:47,440 --> 00:00:52,640

This is basically our only option to give him a better life, otherwise...

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00:00:52,640 --> 00:00:54,120

he'll die.

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00:00:54,120 --> 00:00:57,000

Surgeons are constantly forced to question how far

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00:00:57,000 --> 00:01:00,440

they should push the boundaries in the hope of finding a cure.

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00:01:00,440 --> 00:01:03,840

Sometimes we've got the technology, we can do all kinds of things,

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00:01:03,840 --> 00:01:06,080

but we have to also ask the question,

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00:01:06,080 --> 00:01:07,760

is it the right thing to do?

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00:01:07,760 --> 00:01:10,200

Just because we CAN do it, OUGHT we be doing it?

21

00:01:29,720 --> 00:01:35,160

15-year-old Shauna has spent her entire life in and out of hospital.

22

00:01:35,160 --> 00:01:38,840

She was born with one lung, a major heart defect

23

00:01:38,840 --> 00:01:40,960

and her windpipe is slowly closing.

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00:01:40,960 --> 00:01:44,600

So I have to ring the doorbell now, because I lost my badge.

25

00:01:44,600 --> 00:01:46,480

Do you want to ring the doorbell?

26

00:01:47,680 --> 00:01:50,600

Recently, her ability to breathe has deteriorated

27

00:01:50,600 --> 00:01:53,440

and she is at risk of suffocation.

28

00:01:53,440 --> 00:01:56,960

'They can't give it a name, cos she has that many problems.

29

00:01:56,960 --> 00:01:59,520

'She had a heart attack ten weeks ago...'

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00:01:59,520 --> 00:02:04,200

I nearly lost her that day. It took them seven minutes to get her back.

31

00:02:06,560 --> 00:02:09,880

If they don't do anything pretty quick,

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00:02:09,880 --> 00:02:12,520

I won't have her much longer.

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00:02:12,520 --> 00:02:13,960

They've told me that.

34

00:02:15,200 --> 00:02:21,360

State-of-the-art equipment, huh? You've got a telly in here as well.

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00:02:21,360 --> 00:02:24,720

Shauna is at Great Ormond Street to see if she is eligible for

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00:02:24,720 --> 00:02:28,760

experimental surgery that has only been performed on one other child.

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00:02:28,760 --> 00:02:31,960

Surgeons want to offer her a donor trachea

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00:02:31,960 --> 00:02:36,080

which will be modified with Shauna's own stem cells.

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00:02:36,080 --> 00:02:40,240

If they go ahead, she will be given a new windpipe made of her own DNA.

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00:02:41,320 --> 00:02:43,760

We've only done one tracheal transplant before

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00:02:43,760 --> 00:02:48,040

and we learned a lot of things in a very short period of time

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00:02:48,040 --> 00:02:50,280

and there are no long-term data.

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00:02:50,280 --> 00:02:54,520

The balance between is this appropriate or ethical,

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00:02:54,520 --> 00:02:59,440

right or wrong, is a scenario in which there are no correct answers.

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00:02:59,440 --> 00:03:02,760

There's just best judgement and so that's what we'll try to achieve

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00:03:02,760 --> 00:03:05,840

between us all and get a lot of input into getting to that decision.

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00:03:07,360 --> 00:03:11,200

Colin Wallace is a respiratory consultant at Great Ormond Street.

48

00:03:11,200 --> 00:03:12,560

Hi.

49

00:03:12,560 --> 00:03:16,600

He's part of the team weighing up the risks of the transplant

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00:03:16,600 --> 00:03:18,320

and her current quality of life.

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00:03:18,320 --> 00:03:22,800

Are you Shauna? How're you doing? I've heard lots about you.

52

00:03:22,800 --> 00:03:25,160

- We haven't met before, have we?

- No.

53

00:03:25,160 --> 00:03:31,120

If I was to ask you for three things that you would like to have better

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00:03:31,120 --> 00:03:34,880

after an operation on your trachea, what would they be?

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00:03:34,880 --> 00:03:38,520

There's a lot of things she would like, aren't there?

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00:03:38,520 --> 00:03:42,040

Name one of them - what would you like?

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00:03:42,040 --> 00:03:46,080

- Be better.

- Be better.

- And in what way would you like to be better?

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00:03:48,280 --> 00:03:52,000

What do you miss? You like to do?

59

00:03:52,000 --> 00:03:55,320

- You go with the carers, but you can't do it.

- Swimming.

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00:03:55,320 --> 00:03:57,360

- Swimming.
- Yes?

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00:03:57,360 --> 00:04:02,120

You'd like to try swimming? And you go to school? Normal school?

62

00:04:02,120 --> 00:04:07,640

- Do you have to have someone with you?
- Yes.
- What's that like?
- All right.

63

00:04:07,640 --> 00:04:10,720

- Hm?
- All right.
- It's OK, yes? And at home?

64

00:04:10,720 --> 00:04:12,720

She has night carers.

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00:04:12,720 --> 00:04:16,280

They come in at 8.00 on a night and finish at 6.00 in the morning,

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00:04:16,280 --> 00:04:18,000

so that I can have some rest.

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00:04:18,000 --> 00:04:20,800

Yes. And you have a friend at school?

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00:04:20,800 --> 00:04:23,760

- What's your best friend's name?
- Courtney.

69

00:04:23,760 --> 00:04:26,320

What did she think about your tracheostomy?

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00:04:26,320 --> 00:04:30,800

- She's all right with it.
- She's used to it now, is she?
- Yes.

71

00:04:30,800 --> 00:04:33,920

And what about other things you'd like to do

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00:04:33,920 --> 00:04:35,680

that you can't do at the moment?

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00:04:38,000 --> 00:04:42,560

- Like, on the bus.

- On the bus? You don't go on the bus?

74

00:04:42,560 --> 00:04:47,120

- I do, but...

- But you and Courtney could go by yourselves on the bus.

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00:04:47,120 --> 00:04:52,000

- Yes.

- You think so? Would that be quite an adventure, hey?

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00:04:52,000 --> 00:04:56,440

What would you need to know will happen

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00:04:56,440 --> 00:04:59,120

to be able to go ahead and say yes, we'll have the surgery?

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00:05:01,400 --> 00:05:02,640

Well, it's stupid really,

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00:05:02,640 --> 00:05:05,120

cos I'd want to know if it was going to be a success.

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00:05:05,120 --> 00:05:09,080

- But you can't give me that guarantee.

- Yes.

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00:05:09,080 --> 00:05:11,840

So, because her life is good at the moment,

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00:05:11,840 --> 00:05:13,960

she has got a good quality of life.

83

00:05:13,960 --> 00:05:15,320

Mm, mm.

84

00:05:15,320 --> 00:05:18,320

It's a slightly unpredictable one though, isn't it?

85

00:05:18,320 --> 00:05:20,960

Yes, because of the arrest, ten weeks ago.

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00:05:20,960 --> 00:05:25,320

It shows there's a vulnerability here on the narrowing of the trachea

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00:05:25,320 --> 00:05:29,960

and the lack of reserve of only just having one lung.

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00:05:29,960 --> 00:05:31,440

Yes.

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00:05:34,560 --> 00:05:37,560

Shauna is in charge. You in charge? Yay!

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00:05:39,480 --> 00:05:42,760

What's quite interesting is that mother does perceive

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00:05:42,760 --> 00:05:45,960

the current quality of life as being good.

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00:05:45,960 --> 00:05:50,080

In other words, this is not a situation where we've got

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00:05:50,080 --> 00:05:53,240

nothing to lose by going ahead - they've got quite a bit to lose

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00:05:53,240 --> 00:05:56,720

and this is going to make for a difficult decision.

95

00:05:56,720 --> 00:06:00,280

She's got a reasonable quality of life, she's got her good friend,

96

00:06:00,280 --> 00:06:04,200

she clearly has a sense of humour and enjoying herself -

97

00:06:04,200 --> 00:06:05,600

it makes it harder.

98

00:06:09,360 --> 00:06:13,560

Before surgery becomes an option, doctors examine Shauna's airway

99

00:06:13,560 --> 00:06:17,760

and her lung to see if they are healthy enough to support a transplant.

100

00:06:19,800 --> 00:06:21,640

OK, then?

101

00:06:23,160 --> 00:06:27,000

If you plug it on... Got it? Well done.

102

00:06:27,000 --> 00:06:31,200

I'll turn the juice up slowly - some laughing gas to begin with, OK?

103

00:06:31,200 --> 00:06:33,480

Try and think of something nice.

104

00:06:33,480 --> 00:06:36,960

Some nice place to go when you're asleep.

105

00:06:38,960 --> 00:06:41,200

I think we'll lie you back now, just...

106

00:06:41,200 --> 00:06:43,120

Why don't you lie back gently?

107

00:06:44,400 --> 00:06:47,800

- We'll take care of her.

- See you soon.

- We'll see you later.

108

00:06:47,800 --> 00:06:49,640

- Thanks very much.

- Down we go.

109

00:06:52,520 --> 00:06:54,800

SUCTION

110

00:07:11,800 --> 00:07:15,120

Until now, a metal cage called a stent has been keeping

111

00:07:15,120 --> 00:07:17,160

Shauna's trachea open.

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00:07:17,160 --> 00:07:19,680

Surgeons must adjust it every ten weeks,

113

00:07:19,680 --> 00:07:23,440

but this causes scar tissue to build up and is blocking her airway.

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00:07:25,440 --> 00:07:27,680

It's extraordinary imaging, this -

115

00:07:27,680 --> 00:07:31,080

the trachea has got this metalwork all round it

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00:07:31,080 --> 00:07:34,400

that's supporting it, but inside the trachea, it's very narrow.

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00:07:34,400 --> 00:07:40,640

There's a lot of tissue here that's grown into this stenting process.

118

00:07:40,640 --> 00:07:44,240

It's lying perilously close to very big vessels

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00:07:44,240 --> 00:07:48,520

and then you've got the only lung, which has overblown and

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00:07:48,520 --> 00:07:53,000

extended right across the midline, so it's a huge, single lung.

121

00:07:54,280 --> 00:07:55,640

Encouragingly,

122

00:07:55,640 --> 00:08:00,840

the airways from the trachea onwards are of good calibre

123

00:08:00,840 --> 00:08:03,880

and nicely open.

124

00:08:05,800 --> 00:08:09,640

But my feeling is that on these scans,

125

00:08:09,640 --> 00:08:13,280

we've got enough good lung structure here...

126

00:08:14,360 --> 00:08:16,160

This lung could cope.

127

00:08:22,120 --> 00:08:25,840

As tracheal transplants in children are so new,

128

00:08:25,840 --> 00:08:29,680

surgeon Martin Elliott must consult with the hospital's ethics committee

129

00:08:29,680 --> 00:08:32,000

before offering surgery.

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00:08:32,000 --> 00:08:35,400

It is made up of professionals from a variety of backgrounds,

131

00:08:35,400 --> 00:08:38,640

from both inside and outside the hospital.

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00:08:38,640 --> 00:08:43,920

'It's an extra check on the validity of what we're proposing.'

133

00:08:43,920 --> 00:08:48,600

Frankly, we are so focused on trying to make her better that we

134

00:08:48,600 --> 00:08:54,840

need a more cool and detached intellectual discussion.

135

00:08:54,840 --> 00:09:00,840

I want to show you what we did to another patient last year.

136

00:09:00,840 --> 00:09:05,240

We took a donor trachea and that trachea was then washed

137

00:09:05,240 --> 00:09:07,720

until all the cells were removed from it,

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00:09:07,720 --> 00:09:09,720

so detergent enzymatic washes.

139

00:09:09,720 --> 00:09:12,320

So you're left with what is essentially a scaffold

140

00:09:12,320 --> 00:09:14,960

of largely collagen and just a few proteins,

141

00:09:14,960 --> 00:09:18,360

but no active... immunologically active cells.

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00:09:18,360 --> 00:09:22,440

That graft was marinated with stem cells

143

00:09:22,440 --> 00:09:27,160

and then locally we injected a drug called TGF beta,

144

00:09:27,160 --> 00:09:30,360

which is supposed to trigger it to become cartilage.

145

00:09:30,360 --> 00:09:34,000

So after it's been marinated, the graft returned in a motorbike

146

00:09:34,000 --> 00:09:39,920

to Great Ormond Street and we were able to stitch the trachea into Kieron.

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00:09:39,920 --> 00:09:45,440

So what we want to do for Shauna is to use the same basic principles

148

00:09:45,440 --> 00:09:49,120

of allowing a skeleton of tissue to be populated

149

00:09:49,120 --> 00:09:53,480

by the child's own stem cells, so that there's no rejection,

150

00:09:53,480 --> 00:09:58,160

and to get more rapid cellularisation of the child

151

00:09:58,160 --> 00:10:00,000

than we had time for in Kieron.

152

00:10:00,000 --> 00:10:02,360

I'd just like to know, if you don't do the procedure,

153

00:10:02,360 --> 00:10:05,480

what is likely to be the manner of her death?

154

00:10:05,480 --> 00:10:09,920

What happens as the winter emerges is that the airway,

155

00:10:09,920 --> 00:10:11,960

the trachea is very like your nose,

156

00:10:11,960 --> 00:10:15,360

so as you imagine your nose blocking up when you have a cold,

157

00:10:15,360 --> 00:10:18,360

if you've ever...just remember breathing through a snorkel

158

00:10:18,360 --> 00:10:23,400

for the first time, the sense of not being able to breathe is...

159

00:10:23,400 --> 00:10:26,000

One of the greatest fears that you can have,

160

00:10:26,000 --> 00:10:29,120

so I can't imagine that it would be a pleasant demise.

161

00:10:29,120 --> 00:10:32,680

But she's going to die anyway, even if you do the procedure -

162

00:10:32,680 --> 00:10:35,760

what would be the nature of her death if you've done the procedure?

163

00:10:35,760 --> 00:10:37,120

Um...

164

00:10:38,800 --> 00:10:41,720

Well, I think that's saying more than I'm prepared to say.

165

00:10:41,720 --> 00:10:45,960

I don't think she will die if we do the procedure. Um...

166

00:10:45,960 --> 00:10:49,880

We're trying to do the procedure so that she doesn't.

167

00:10:49,880 --> 00:10:51,800

Or at least we prolong her life

168

00:10:51,800 --> 00:10:55,160

and her quality of life for as long as possible.

169

00:10:55,160 --> 00:10:58,560

In terms of the timing of the decision-making,

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00:10:58,560 --> 00:11:00,960

would there be any advantages in deferring?

171

00:11:00,960 --> 00:11:03,280

Would it give better outcomes or whatever?

172

00:11:03,280 --> 00:11:06,800

No, I think there'd be a disadvantage to deferment.

173

00:11:06,800 --> 00:11:09,720

The section here is only 2mm across.

174

00:11:09,720 --> 00:11:12,000

That's the tip of a Biro.

175

00:11:12,000 --> 00:11:15,680

At that point, you have to really consider how much

176

00:11:15,680 --> 00:11:18,000
swelling of the mucosa you can tolerate

177

00:11:18,000 --> 00:11:20,440
before it becomes a critical narrowing.

178

00:11:20,440 --> 00:11:23,040
You haven't mentioned palliative care as an option.

179

00:11:23,040 --> 00:11:26,560
Is that because you think it's inappropriate?

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00:11:26,560 --> 00:11:29,480
No, I think if I thought the treatment was really horrific,

181

00:11:29,480 --> 00:11:31,760
and we hadn't got the experience that we have had,

182

00:11:31,760 --> 00:11:34,440
I wouldn't have any hesitation in offering that.

183

00:11:34,440 --> 00:11:37,880
I don't not offer palliative care for complex patients.

184

00:11:37,880 --> 00:11:41,600
So it's not really just a choice between a horrible death

185

00:11:41,600 --> 00:11:44,400
and the chance of improved quality of life?

186

00:11:44,400 --> 00:11:47,800
There is a third option, which is a less horrible death?

187

00:11:49,040 --> 00:11:50,880
Yes, that's true.

188

00:11:50,880 --> 00:11:53,560
Palliative care may still become necessary,

189

00:11:53,560 --> 00:11:56,560
but at the moment I'm not sure that the technical solution

190

00:11:56,560 --> 00:12:01,400
does not indeed trump the palliative care.

191

00:12:01,400 --> 00:12:05,040

But only for the willing participant in uncertainty.

192

00:12:12,600 --> 00:12:14,520

Do we have any further questions?

193

00:12:16,960 --> 00:12:19,800

The committee debated for another hour

194

00:12:19,800 --> 00:12:22,760

and found no ethical objections to Shauna's surgery.

195

00:12:26,800 --> 00:12:28,760

Shauna will return to Middlesbrough

196

00:12:28,760 --> 00:12:30,960

and may have to wait up to four months

197

00:12:30,960 --> 00:12:32,720

for the new trachea to be grown.

198

00:12:51,200 --> 00:12:54,400

Three-month-old Muhammed has just arrived

199

00:12:54,400 --> 00:12:57,040

on the cardiac intensive care unit.

200

00:12:57,040 --> 00:13:00,160

At birth, he was diagnosed with Jeune's Syndrome,

201

00:13:00,160 --> 00:13:06,640

a very rare condition with only 125 recorded cases worldwide.

202

00:13:06,640 --> 00:13:08,760

Muhammed's rib cage doesn't grow.

203

00:13:08,760 --> 00:13:11,920

It is trapping his lungs and eventually,

204

00:13:11,920 --> 00:13:15,680

he will be unable to produce enough oxygen to stay alive.

205

00:13:16,880 --> 00:13:19,440

Can we drop these lights now?

206

00:13:19,440 --> 00:13:22,320

The width of the abdomen is normal width for a child

207

00:13:22,320 --> 00:13:26,960

and if you just imagine where the soft tissue ends on the right with the vague,

208

00:13:26,960 --> 00:13:30,040

grey things on either side, that would be the normal width.

209

00:13:30,040 --> 00:13:31,600

So that's baby size.

210

00:13:31,600 --> 00:13:33,400

If you come up towards the chest,

211

00:13:33,400 --> 00:13:37,800

the chest narrows right in and then comes out again in the armpits.

212

00:13:37,800 --> 00:13:40,760

This is characteristic of Jeune's Syndrome.

213

00:13:40,760 --> 00:13:43,800

So this chest here is squeezing in

214

00:13:43,800 --> 00:13:49,360

and the heart in the middle and the lungs are actually squashed,

215

00:13:49,360 --> 00:13:52,760

so the child's chest is going to stay the same size.

216

00:13:52,760 --> 00:13:57,280

So as the baby gets bigger, the chest doesn't grow and the lungs get smashed up.

217

00:13:58,280 --> 00:14:03,600

Muhammed's parents were told by their hospital in Birmingham that they could offer no treatment.

218

00:14:03,600 --> 00:14:07,080

After searching on the Internet, Muhammed's father

219

00:14:07,080 --> 00:14:10,400

read about an experimental trial at Great Ormond Street,

220

00:14:10,400 --> 00:14:15,440

where they expand the rib cage in the hope that the lungs will grow.

221

00:14:16,720 --> 00:14:21,240

Already approved by the ethics committee, it's the only treatment for Jeune's in Europe

222

00:14:21,240 --> 00:14:25,800

and has been performed on just six children before now.

223

00:14:25,800 --> 00:14:29,040

- Hello, hi.

- Hi.

- My name is Martin Elliott.

224

00:14:29,040 --> 00:14:31,280

Hi, very nice to meet you.

225

00:14:31,280 --> 00:14:34,600

- Martin Elliott. Neil Bulstrode.

- Nice to meet you.

226

00:14:34,600 --> 00:14:40,080

The operation is really very simple and the principle is simple,

227

00:14:40,080 --> 00:14:42,840

so we're going to try and show you what it is - we've worked out a way

228

00:14:42,840 --> 00:14:45,000

of doing this using Neil's hands.

229

00:14:45,000 --> 00:14:50,040

we cut the ribs in the front and the back and bring them out

230

00:14:50,040 --> 00:14:52,760

and reconnect them...

231

00:14:52,760 --> 00:14:55,360

with plates and screws,

232

00:14:55,360 --> 00:14:57,200

so effectively,

233

00:14:57,200 --> 00:15:02,840

we're going from four ribs, to two ribs, which are longer.

234

00:15:02,840 --> 00:15:07,280

We would cut the third one in the middle,

235

00:15:07,280 --> 00:15:11,280

then we would cut the other ones, front and back, front and back,

236

00:15:11,280 --> 00:15:13,720

so that we could make them longer

237

00:15:13,720 --> 00:15:17,720

but leave the third and the bottom one loose.

238

00:15:17,720 --> 00:15:20,160

So, it's just stretching it out.

239

00:15:20,160 --> 00:15:23,360

And at some stage in the future we can do the next stage

240

00:15:23,360 --> 00:15:25,360

and make his chest wall even bigger.

241

00:15:26,440 --> 00:15:30,480

We've done a very small number of patients with this condition

242

00:15:30,480 --> 00:15:35,960

in England and they've all got back home off a ventilator.

243

00:15:35,960 --> 00:15:37,680

All got back home.

244

00:15:37,680 --> 00:15:41,800

One of them has sadly died later

245

00:15:41,800 --> 00:15:45,280

and we don't know what's going to happen to the others.

246

00:15:45,280 --> 00:15:49,320

No-one's going to force you to do something to Muhammed like this,

247

00:15:49,320 --> 00:15:51,600

which is so uncertain.

248

00:15:52,920 --> 00:15:58,680

Erm...if you sign a consent form, you're basically signing

249

00:15:58,680 --> 00:16:00,920

a contract with uncertainty.

250

00:16:00,920 --> 00:16:04,600

We don't know what is going to happen and neither can you.

251

00:16:04,600 --> 00:16:08,360

If it does go well and he needs surgeries in the future,

252

00:16:08,360 --> 00:16:11,560

how often would he need surgeries?

253

00:16:11,560 --> 00:16:14,080

Well, the follow-up would be lifelong.

254

00:16:14,080 --> 00:16:15,640

We will always keep a close eye.

255

00:16:15,640 --> 00:16:18,440

People in Birmingham will keep a close eye on him

256

00:16:18,440 --> 00:16:22,160

and then phone us when things change. That's what we would expect.

257

00:16:23,560 --> 00:16:26,800

Basically, the way I feel is...

258

00:16:28,480 --> 00:16:31,640

We've got no other option, basically.

259

00:16:31,640 --> 00:16:35,920

Even, whatever happens, erm...

260

00:16:37,960 --> 00:16:40,120

..I think, we both need to give it a try.

261

00:16:40,120 --> 00:16:41,360

Yeah.

262

00:16:41,360 --> 00:16:44,000

Yeah. Are you sure you're happy with that too?

263

00:16:45,560 --> 00:16:49,880

We don't want to cause Muhammed or you any suffering.

264

00:16:49,880 --> 00:16:54,320

That's not what we're trying to do. But that might happen.

265

00:16:56,000 --> 00:16:59,760

At least in our minds, we will know we have tried.

266

00:16:59,760 --> 00:17:01,520

What we can do as parents.

267

00:17:03,200 --> 00:17:05,880

I think that's the main thing.

268

00:17:05,880 --> 00:17:08,360

OK, well, we'll do our best as well.

269

00:17:08,360 --> 00:17:10,320

And...

270

00:17:10,320 --> 00:17:12,880

we'd look after him as though he was our own

271

00:17:12,880 --> 00:17:14,960

and make sure he's all right.

272

00:17:24,560 --> 00:17:26,880

It is not compulsory to have treatment for this

273

00:17:26,880 --> 00:17:30,320

because no-one knows what the future is.

274

00:17:30,320 --> 00:17:34,520

We certainly wouldn't pressurise a family into doing this.

275

00:17:34,520 --> 00:17:37,520

Absolutely the opposite, we would very much explain to them -

276

00:17:37,520 --> 00:17:41,720

and have explained to them - that this wouldn't be the only choice.

277

00:17:43,480 --> 00:17:47,440

The option is to accept that death is inevitable.

278

00:17:47,440 --> 00:17:50,800

I know the Birmingham team have even put that in a more explicit way

279

00:17:50,800 --> 00:17:53,400

because they said, "We wouldn't do this."

280

00:17:53,400 --> 00:17:56,480

And a significant proportion of doctors

281

00:17:56,480 --> 00:17:59,920

and parents around the world would reasonably take that option.

282

00:17:59,920 --> 00:18:05,200

But the family have been much more on the side of giving it a whirl

283

00:18:03,080 --> 00:18:05,200

to see if we can help.

284

00:18:19,000 --> 00:18:21,960

24 hours later and Muhammed is ready for surgery.

285

00:18:21,960 --> 00:18:26,000

Until surgeons open him up and see the state of his ribs,

286

00:18:26,000 --> 00:18:29,560

they won't know exactly how many they can expand.

287

00:19:17,440 --> 00:19:18,600

Big vessel.

288

00:19:18,600 --> 00:19:22,800

- We cut four at the back.

- Four at the back.

289

00:19:22,800 --> 00:19:25,840

Five at the front, that's what we've done before.

290

00:19:25,840 --> 00:19:28,680

But it doesn't really matter where we start, does it?

291

00:19:28,680 --> 00:19:31,680

So I'm just going to move a little bit down towards you.

292

00:19:31,680 --> 00:19:33,200

May I have a blade on here now?

293

00:19:33,200 --> 00:19:35,600

Let's find out where we're going to cut across here.

294

00:19:35,600 --> 00:19:37,760
- We think there, probably, don't we?
- Yep.

295
00:19:41,160 --> 00:19:42,320
Down to eight.

296
00:19:43,760 --> 00:19:46,240
There won't be much room, will there?

297
00:19:47,520 --> 00:19:49,560
Gosh, it's tight.

298
00:19:49,560 --> 00:19:53,480
It's amazing they can breathe at all when you put so much force on here.

299
00:19:55,160 --> 00:19:56,280
Diathermy, please.

300
00:19:56,280 --> 00:19:57,560
Go ahead.

301
00:19:58,760 --> 00:20:01,160
So, we've just, having cut those ribs,

302
00:20:01,160 --> 00:20:06,800
and now we've brought them forwards, so this is one rib up

303
00:20:06,800 --> 00:20:12,680
and one rib down, joined together, so if I let it go, it forms an arch.

304
00:20:12,680 --> 00:20:16,680
So it's lifted the whole chest wall by a good two centimetres.

305
00:20:18,680 --> 00:20:21,840
Once Neil Bulstrode can see the expanded ribs,

306
00:20:21,840 --> 00:20:26,360
he can then shape the titanium plates which will hold them together.

307
00:20:32,240 --> 00:20:36,080
That's good. That's going to be great. OK, the clamps, please.

308
00:20:36,080 --> 00:20:39,960
That one, yep. And we need a syringe of saline.

309

00:20:50,880 --> 00:20:54,800

The drill didn't go all the way through the...

310

00:20:54,800 --> 00:20:56,720

This is the eight, please.

311

00:20:56,720 --> 00:20:58,840

DRILL WHIRRS

312

00:21:13,520 --> 00:21:15,560

Eight, please.

313

00:21:16,560 --> 00:21:18,840

Great.

314

00:21:18,840 --> 00:21:20,880

Just check for any...

315

00:21:20,880 --> 00:21:24,040

The titanium plate has screwed in beautifully.

316

00:21:25,440 --> 00:21:31,240

So you can see now, with the four ribs cut in a staggered fashion,

317

00:21:31,240 --> 00:21:35,280

they've now been rejoined to lengthen the rib.

318

00:21:36,600 --> 00:21:40,960

I'd like to do the other ribs, but these are too small. You can't expand everything.

319

00:21:40,960 --> 00:21:43,560

So we get them to stage two and then,

320

00:21:43,560 --> 00:21:45,760

yeah, later on we come back and do the lower ribs

321

00:21:45,760 --> 00:21:48,080

through a separate incision and see how we get on.

322

00:21:48,080 --> 00:21:50,160

And the top ones, we may never be able to do.

323

00:21:51,520 --> 00:21:53,160

I think that's all right.

324

00:21:53,160 --> 00:21:56,480

- If we just stitch the other stuff up, it'll be OK, won't it?
- Yeah, yeah.

325

00:22:03,960 --> 00:22:06,920

It has taken one and a half hours to break and expand

326

00:22:06,920 --> 00:22:09,120

the left side of Muhammed's rib cage.

327

00:22:09,120 --> 00:22:12,520

They now have to do the same to the right.

328

00:22:13,920 --> 00:22:17,840

The ribs are very short and small, as you can perhaps see,

329

00:22:17,840 --> 00:22:21,280

so there's not really very much space and it's also rather fragile,

330

00:22:21,280 --> 00:22:25,120

so we basically want to get it right first time.

331

00:22:25,120 --> 00:22:29,720

But it went very well and as soon as we had enlarged both those ribs,

332

00:22:29,720 --> 00:22:32,480

Anne, the anaesthetist, said that the ventilation

333

00:22:32,480 --> 00:22:34,600

had got enormously better instantly.

334

00:22:34,600 --> 00:22:38,560

So when we double that effect by doing the other side,

335

00:22:38,560 --> 00:22:41,800

it should be greatly improved.

336

00:22:49,440 --> 00:22:51,600

After three hours of surgery,

337

00:22:51,600 --> 00:22:54,040

Muhammed is taken back to intensive care.

338

00:22:55,240 --> 00:22:59,760

He will remain on a ventilator and sedated for the next few days.

339

00:22:59,760 --> 00:23:02,600

His lungs will be monitored round-the-clock

340

00:23:02,600 --> 00:23:05,400

to see if they adapt to his expanded rib cage.

341

00:23:07,960 --> 00:23:11,520

The expansion on both sides went according to plan

342

00:23:11,520 --> 00:23:15,600

and immediately the anaesthetist said there was an improvement

343

00:23:15,600 --> 00:23:18,480

in the ability to ventilate Muhammed,

344

00:23:18,480 --> 00:23:21,760

- so we were really pleased with that.

- Yeah, yeah.

345

00:23:21,760 --> 00:23:25,200

- He's doing really well.

- That's a really good news, yeah.

346

00:23:25,200 --> 00:23:27,880

- I was really concerned.

- Of course.

347

00:23:27,880 --> 00:23:32,280

- But thank God that's gone good, yeah?

- Exactly. Exactly.

348

00:23:32,280 --> 00:23:34,320

Do you have any other questions?

349

00:23:34,320 --> 00:23:36,440

SHE SPEAKS IN OWN LANGUAGE

350

00:23:38,360 --> 00:23:42,520

Yeah, when we tried feeding him with a bottle, he wouldn't drink before.

351

00:23:42,520 --> 00:23:46,880

- He was coughing a lot.

- Right.

- Would he still have the same issues?

352

00:23:46,880 --> 00:23:50,160

We'll have to see exactly how that goes.

353

00:23:50,160 --> 00:23:53,680

Sometimes we can put another little tube in through his nose

354

00:23:53,680 --> 00:23:56,360

which goes into the stomach and, if necessary,

355

00:23:56,360 --> 00:23:59,400

we can feed him through there temporarily.

356

00:24:12,080 --> 00:24:14,920

- You can tell the difference with the chest already.

- Chest, yeah.

357

00:24:14,920 --> 00:24:17,760

You can already see the increase in the size.

358

00:24:20,600 --> 00:24:24,320

And now we just want his lungs to grow into that extra space.

359

00:24:56,280 --> 00:24:58,160

Three days after surgery,

360

00:24:58,160 --> 00:25:01,800

Muhammad's lungs are struggling to expand in his new rib cage.

361

00:25:03,200 --> 00:25:06,800

The ventilation's still not perfect, you know, so there's still

362

00:25:06,800 --> 00:25:12,080

some episodes where the SATs fall and some episodes where it's a bit hard to get air into the lungs,

363

00:25:12,080 --> 00:25:15,360

and also, I guess, he's not quite moving as much air yet

364

00:25:15,360 --> 00:25:19,400

as we thought, but it's only a few days since he had a big operation

365

00:25:19,400 --> 00:25:21,400

and we broke quite a lot of ribs,

366

00:25:21,400 --> 00:25:25,080

so if you can imagine what that would feel like for you or I -

367

00:25:25,080 --> 00:25:28,320

we probably wouldn't want to move much air anyway.

368

00:25:28,320 --> 00:25:31,080

But is that a concern?

369

00:25:31,080 --> 00:25:33,760

I think we are still within the window

370

00:25:33,760 --> 00:25:36,200

of what we, kind of, expect after this.

371

00:25:36,200 --> 00:25:39,040

It does take a while for the lung to spring open

372

00:25:39,040 --> 00:25:41,840

and get used to that new way of having to work.

373

00:25:41,840 --> 00:25:46,120

I think more time has to go by before we can say for sure

374

00:25:46,120 --> 00:25:49,680

how much of a success it's been and what happens now.

375

00:25:53,760 --> 00:25:56,920

What families in this position have to get to grips with

376

00:25:56,920 --> 00:25:59,600

is that they are part of an experiment

377

00:25:59,600 --> 00:26:02,120

and that we don't know enough to be able to give

378

00:26:02,120 --> 00:26:05,560

the kind of reassurance that we'd be able to give to someone

379

00:26:05,560 --> 00:26:08,800

if we done 100 or 200 or 400 operations which were always the same.

380

00:26:08,800 --> 00:26:11,760

Here we have to make modifications each time we do it

381

00:26:11,760 --> 00:26:16,560

based on what we learned last time and what science has been going on in the background elsewhere.

382

00:26:16,560 --> 00:26:22,920

Everything is subtly different and I can't really give guarantees.

383

00:26:22,920 --> 00:26:26,600

What we have to have is an understanding of uncertainty.

384

00:26:33,720 --> 00:26:39,040

A week after his surgery, Muhammed is transferred back to his Birmingham hospital to recover.

385

00:26:43,840 --> 00:26:47,640

Three months later, his lungs are showing no signs of growth

386

00:26:47,640 --> 00:26:51,120

and his breathing is becoming increasingly difficult.

387

00:26:53,920 --> 00:26:56,800

We've probably got to the end of the road

388

00:26:56,800 --> 00:26:59,040

as far as what we can do for him.

389

00:26:59,040 --> 00:27:02,200

I don't think there's any point in putting him through

390

00:27:02,200 --> 00:27:05,440

another operation to enlarge his chest in any other way

391

00:27:05,440 --> 00:27:08,560

because the lungs have had a trial of survival and failed it.

392

00:27:10,760 --> 00:27:14,040

Doctors in Birmingham have told Muhammed's father

393

00:27:14,040 --> 00:27:16,520

they want to take him off life support.

394

00:27:16,520 --> 00:27:20,240

He has asked to speak to Martin Elliott before he can agree

395

00:27:20,240 --> 00:27:23,200

and a conference call has been arranged.

396

00:27:24,480 --> 00:27:28,160

It's very difficult to know how to help the family through this.

397

00:27:28,160 --> 00:27:30,880

There isn't a way.

398

00:27:30,880 --> 00:27:33,040

It's just painful.

399

00:27:35,120 --> 00:27:37,560

- Um, hi, Professor.

- Hello.

400

00:27:40,880 --> 00:27:41,960

Yeah.

401

00:27:49,880 --> 00:27:52,000

Well, um, what I said to you was

402

00:27:52,000 --> 00:27:55,440

I didn't know whether the lungs would grow at all

403

00:27:55,440 --> 00:27:58,200

and, to be certain that there's going to be some improvement,

404

00:27:58,200 --> 00:28:02,200

you need a long period of time, but if there's been deterioration in between times

405

00:28:02,200 --> 00:28:05,000

I think that really gives us the answer

406

00:28:05,000 --> 00:28:08,040

that there just simply isn't enough lung

407

00:28:08,040 --> 00:28:11,520

for Muhammed to be able to survive on his own.

408

00:28:13,200 --> 00:28:15,240

OK.

409

00:28:15,240 --> 00:28:16,800

OK.

410

00:29:24,040 --> 00:29:28,920

I'm obviously very sorry that we haven't been able to do more for Muhammed and...

411

00:29:43,160 --> 00:29:44,400
Yeah.

412

00:29:46,760 --> 00:29:48,280
OK, well...

413

00:29:49,320 --> 00:29:53,080
We all send you our best wishes from here.

414

00:29:53,080 --> 00:29:54,640
- Thank you.
- Thank you.

415

00:29:54,640 --> 00:29:58,000
- Bye.
- Thanks a lot. Cheers. Bye.

416

00:30:00,680 --> 00:30:03,120
HE SIGHS

417

00:30:04,880 --> 00:30:06,360
OK.

418

00:30:12,840 --> 00:30:15,240
Would we do it again?

419

00:30:15,240 --> 00:30:19,360
For the time being, if we accumulate enough patients in this category

420

00:30:19,360 --> 00:30:21,720
to say, "Look, this is never going to work.

421

00:30:21,720 --> 00:30:27,920
"This category of patients are inappropriate for this therapy because of X or Y",

422

00:30:27,920 --> 00:30:31,240
then clearly that would be relevant, but I don't think we're there yet.

423

00:30:31,240 --> 00:30:34,080
We're still trying to find out what X and Y are

424

00:30:34,080 --> 00:30:36,360
that would stop you doing something.

425

00:30:42,880 --> 00:30:46,080

A few months later and another case of Jeune's syndrome

426

00:30:46,080 --> 00:30:49,480

has been transferred from Stepping Hill Hospital to Great Ormond Street.

427

00:30:50,720 --> 00:30:53,680

Joshua Burns Adair, he's five months old now

428

00:30:53,680 --> 00:30:56,920

and has come down for another expansion tomorrow.

429

00:30:56,920 --> 00:31:00,360

He's got Jeune's thoracic dystrophy, a chronic lung disease.

430

00:31:00,360 --> 00:31:02,760

been up to a maximum of 90% oxygen

431

00:31:02,760 --> 00:31:05,560

and he was in 45% when we picked him up.

432

00:31:05,560 --> 00:31:08,160

That's about that, really, isn't it?

433

00:31:10,200 --> 00:31:13,800

Joshua's condition is critical and despite the dangers,

434

00:31:13,800 --> 00:31:16,880

his parents are eager for the chest expansion.

435

00:31:16,880 --> 00:31:19,120

Surgery is scheduled for tomorrow morning.

436

00:31:20,520 --> 00:31:22,400

After doing everything on the internet,

437

00:31:22,400 --> 00:31:26,440

I brought it up with the doctors and I pushed them to look into it

438

00:31:26,440 --> 00:31:30,880

and it was via, basically, our consultant finding out

439

00:31:30,880 --> 00:31:34,040

that there was actually a surgeon down here

440

00:31:34,040 --> 00:31:36,120
that can do this operation.

441

00:31:36,120 --> 00:31:40,400
- We decided that we wanted to go down every avenue, didn't we?
- Yeah.

442

00:31:40,400 --> 00:31:44,040
To see that we knew if anything did happen to Joshua,

443

00:31:44,040 --> 00:31:46,320
we'd done everything we could.

444

00:31:46,320 --> 00:31:51,400
So this is basically our only option to give him a better life,

445

00:31:51,400 --> 00:31:55,000
otherwise...he'll die.

446

00:31:55,000 --> 00:31:57,560
ALARM WAILS

447

00:31:57,560 --> 00:32:02,040
Whilst Joshua is being transferred from the ambulance ventilator

448

00:32:02,040 --> 00:32:05,520
to one on intensive care, his heart stops beating.

449

00:32:05,520 --> 00:32:07,960
It takes two minutes to revive him.

450

00:32:10,760 --> 00:32:12,440
The operation is cancelled

451

00:32:12,440 --> 00:32:16,160
and his parents are left waiting for the surgery to be rescheduled.

452

00:32:19,400 --> 00:32:23,720
The way they've been speaking for the last month when we had telephone
conversations,

453

00:32:23,720 --> 00:32:27,000
their hopes are very high we can fix him, even though

454

00:32:27,000 --> 00:32:30,200
they have been spoken to and they know that this is experimental.

455

00:32:30,200 --> 00:32:33,600

But Joshua is different now than he was two days ago and I think

456

00:32:33,600 --> 00:32:38,280

if he doesn't get back to his transfer settings

457

00:32:38,280 --> 00:32:43,960

then the surgery is not an option and we have to go down that avenue.

458

00:32:43,960 --> 00:32:47,000

The issue for us and the reason that we're isolated

459

00:32:47,000 --> 00:32:49,160

is that the CO2 is all over the place

460

00:32:49,160 --> 00:32:51,760

and ultimately that's not good for the lungs

461

00:32:51,760 --> 00:32:54,880

and it's not going to be good for multiple procedures

462

00:32:54,880 --> 00:32:59,640

so I would think it would be daft to operate in the next day.

463

00:32:59,640 --> 00:33:01,480

Yeah, I mean, I would just say do we,

464

00:33:01,480 --> 00:33:03,840

after we've got through this next 48 hours,

465

00:33:03,840 --> 00:33:06,160

see if we can optimise him as much as we can

466

00:33:06,160 --> 00:33:08,080

and then we know where we are.

467

00:33:08,080 --> 00:33:12,000

We should then reappraise whether we should actually offer surgery or not.

468

00:33:12,000 --> 00:33:14,040

I think we just need to wait until we've got that.

469

00:33:14,040 --> 00:33:16,440

When he's ready for a general anaesthetic.

470

00:33:16,440 --> 00:33:18,720

I'm not sure what we can do at this stage.

471

00:33:26,240 --> 00:33:32,320

He's at the same ventilation as when he came over to us here.

472

00:33:32,320 --> 00:33:37,720

If he can achieve that then there's a possibility that he could do the surgery,

473

00:33:37,720 --> 00:33:41,200

but if not, surgery may not be his best option.

474

00:33:44,840 --> 00:33:46,600

Yeah.

475

00:33:50,400 --> 00:33:54,000

So what would that mean then? It would just be a matter of him...

476

00:33:56,960 --> 00:34:02,120

- Being left to his own, sort of, agenda, as such?

- Yes.

- Until they...?

477

00:34:02,120 --> 00:34:05,560

Until, yes, he can't sustain himself any more.

478

00:34:13,760 --> 00:34:16,360

You know, whenever you have any patient,

479

00:34:16,360 --> 00:34:18,800

it's trying to make the right decision

480

00:34:18,800 --> 00:34:21,920

and when they are so severe...

481

00:34:23,360 --> 00:34:27,520

..it... Possibly the right decision is not to operate on them

482

00:34:27,520 --> 00:34:31,520

and I think if Joshua stays in his current state

483

00:34:31,520 --> 00:34:34,640

that that'll be our answer.

484

00:34:34,640 --> 00:34:37,640
However, we're all optimistic people

485
00:34:37,640 --> 00:34:40,680
that want to do things to try and help,

486
00:34:40,680 --> 00:34:43,920
but sometimes we have to step back

487
00:34:43,920 --> 00:34:47,680
and admit that maybe we cannot help them.

488
00:34:47,680 --> 00:34:50,440
Always wore a red cloak with a warm hood

489
00:34:50,440 --> 00:34:53,600
and so she was called Little Red Riding Hood.

490
00:34:53,600 --> 00:34:56,560
One day she decided to visit her granny who lived some way

491
00:34:56,560 --> 00:34:58,560
from the woodcutter's cottage.

492
00:34:58,560 --> 00:35:02,920
She took a basket with a cake her mother had baked and set off.

493
00:35:02,920 --> 00:35:06,520
Now, the last thing her mother had said to Little Red Riding Hood was,

494
00:35:06,520 --> 00:35:10,680
"Don't leave the path and don't talk to any strangers."

495
00:35:10,680 --> 00:35:14,200
I'm afraid Little Red Riding Hood was not really listening.

496
00:35:21,720 --> 00:35:25,480
48 hours later and Joshua's oxygen levels have improved.

497
00:35:29,720 --> 00:35:32,200
I guess the key discussion for us is

498
00:35:32,200 --> 00:35:36,000
he's improved, has he improved to the point we can do surgery?

499

00:35:36,000 --> 00:35:40,120

Is this our window for surgery and if we wait are going to miss that?

500

00:35:40,120 --> 00:35:45,480

I am not convinced that there's potential to improve an awful lot further,

501

00:35:45,480 --> 00:35:48,760

so if we are going to wait a few more days,

502

00:35:48,760 --> 00:35:51,560

it's not clear to me exactly what we are waiting for,

503

00:35:51,560 --> 00:35:53,720

because things may not get much better.

504

00:35:53,720 --> 00:35:56,440

I completely agree that this may be the window

505

00:35:56,440 --> 00:35:57,760

that we are actually getting

506

00:35:57,760 --> 00:35:59,800

for treating Joshua in the right direction,

507

00:35:59,800 --> 00:36:02,240

but if anything can be done, probably,

508

00:36:02,240 --> 00:36:05,680

this is the window we need to look at for doing this expansion.

509

00:36:05,680 --> 00:36:09,840

I think, you know, we would have to have some serious discussion

510

00:36:09,840 --> 00:36:14,200

with the family that the risk is he could die on the table, couldn't he?

511

00:36:14,200 --> 00:36:16,920

That is a very real possibility as well.

512

00:36:16,920 --> 00:36:19,800

You know, we should continue to also say

513

00:36:19,800 --> 00:36:23,080

that if they felt that they didn't want to go down this route

514

00:36:23,080 --> 00:36:27,440
given the risks now that we would fully support them in that decision as well.

515
00:36:27,440 --> 00:36:32,920
They have to know that he has about a one in two chance of coming through.

516
00:36:32,920 --> 00:36:35,280
You're semi-detached from this, Ruth.

517
00:36:35,280 --> 00:36:37,680
- Well, I am.
- Are you disturbed by it?
- Yes, to be honest.

518
00:36:37,680 --> 00:36:42,080
I've just come into this and don't really know the case,

519
00:36:42,080 --> 00:36:46,360
but I have to say I am kind of taken aback about going forward

520
00:36:46,360 --> 00:36:49,520
for such dramatic surgery with a risk of 50%.

521
00:36:49,520 --> 00:36:53,240
That's... And I am detached. You know, I don't know the child.

522
00:36:53,240 --> 00:36:58,320
It's a good point. On the other side of the coin is, sort of,

523
00:36:58,320 --> 00:37:02,520
the alternative is 100% chance of death within a year probably.

524
00:37:03,760 --> 00:37:06,800
I'd be very surprised if this child was alive in six months, even,

525
00:37:06,800 --> 00:37:09,200
if he didn't have surgery.

526
00:37:09,200 --> 00:37:15,480
And so that's the... The risk is in that context.

527
00:37:15,480 --> 00:37:21,080
That, sort of, to me... On the one hand, is this whole thing crazy?

528
00:37:22,280 --> 00:37:27,720
This is right at the edge of what I feel we're fully comfortable with doing.

529

00:37:27,720 --> 00:37:30,000

Really right at the edge of it and...

530

00:37:31,200 --> 00:37:34,960

I've brought Ruth, our trainee, along today - it's clear you feel exactly the same.

531

00:37:34,960 --> 00:37:37,560

Or maybe it's over your edge. So, I'll be honest about it,

532

00:37:37,560 --> 00:37:40,680

but as long as the parents are absolutely clear

533

00:37:40,680 --> 00:37:43,360

that this is as far as, ethically, we feel able to go

534

00:37:43,360 --> 00:37:48,360

and they understand how big the risks are, then...well, OK.

535

00:37:53,760 --> 00:37:58,600

In the last 24 hours, we've turned the ventilator down some more

536

00:37:58,600 --> 00:38:01,920

and so that's quite a lot more encouraging

537

00:38:01,920 --> 00:38:05,640

and I personally feel a lot more comforted seeing that coming down.

538

00:38:05,640 --> 00:38:07,920

Now, that doesn't by any means mean

539

00:38:07,920 --> 00:38:13,200

that means we're going to sail through this procedure, but I think...

540

00:38:13,200 --> 00:38:15,600

I get the feeling we're at the level,

541

00:38:15,600 --> 00:38:17,720

the best we could have hoped for

542

00:38:17,720 --> 00:38:20,760

So I think we've got to a point

543

00:38:20,760 --> 00:38:22,880

where if we're going to do it,

544

00:38:22,880 --> 00:38:25,640
probably now is the best time we can.

545

00:38:26,920 --> 00:38:29,120
There are some risks that we do not know.

546

00:38:29,120 --> 00:38:32,440
As I said, the experience on the surgery's quite dangerous.

547

00:38:32,440 --> 00:38:35,680
The known risks are there are chances which are very high

548

00:38:35,680 --> 00:38:40,120
that he will not come through, or he will not make it after surgery alive.

549

00:38:40,120 --> 00:38:42,800
Other risks, which we know definitely,

550

00:38:42,800 --> 00:38:45,640
are Joshua will need multiple surgeries

551

00:38:45,640 --> 00:38:50,080
and at any stage he may fail to cope with all these interventions. We need to be aware of that.

552

00:38:51,280 --> 00:38:55,040
Our own experience, we have lost two children.

553

00:38:55,040 --> 00:38:59,760
When we say 50%, what we're saying is it's very high, you know?

554

00:38:59,760 --> 00:39:03,480
It means if you took two children into the operating theatre,

555

00:39:03,480 --> 00:39:06,240
only one of the children would come back out,

556

00:39:06,240 --> 00:39:11,200
but the other thing that people felt we should also just discuss

557

00:39:11,200 --> 00:39:14,080
is if things sadly go wrong in theatre,

558

00:39:14,080 --> 00:39:17,560

would you want to come into theatre at that point?

559

00:39:17,560 --> 00:39:20,680

We would talk to you at that time. But...

560

00:39:20,680 --> 00:39:23,640

I'd rather... I don't want to see... I mean, obviously...

561

00:39:23,640 --> 00:39:27,280

Well, I don't know. I don't know. Ask me that question, "If."

562

00:39:27,280 --> 00:39:30,040

It's one to think about. I probably would.

563

00:39:30,040 --> 00:39:32,120

Yeah, I don't know. I can't say.

564

00:39:32,120 --> 00:39:34,200

On one hand, I don't want to take you there

565

00:39:34,200 --> 00:39:36,600

because we hope we're going to get through,

566

00:39:36,600 --> 00:39:39,320

but it's just something to think about.

567

00:39:39,320 --> 00:39:43,160

All I can say... We'll cross that bridge if we come to it.

568

00:39:43,160 --> 00:39:45,160

Yeah. Fair enough.

569

00:39:56,800 --> 00:40:01,240

It was extremely difficult for me to put this in words to them, but it's my duty to.

570

00:40:01,240 --> 00:40:05,040

You'll really be able to transfer your humane abilities

571

00:40:05,040 --> 00:40:08,400

in the discussion process, convey them to parents,

572

00:40:08,400 --> 00:40:11,280

help them to make a decision, but once the decision's made,

573

00:40:11,280 --> 00:40:13,920
you should be strong enough to carry on.

574
00:40:13,920 --> 00:40:15,680
There's no backing out.

575
00:40:21,440 --> 00:40:24,280
My brave little soldier. Aren't you?

576
00:40:24,280 --> 00:40:27,360
You know, to your mother, you're everything in the world.

577
00:40:32,960 --> 00:40:37,160
You show them. Yeah? You show 'em.

578
00:40:44,720 --> 00:40:47,360
This is Joshua Burns Adair.

579
00:40:47,360 --> 00:40:51,960
Consented for lateral chest-expansion on both sides with metallic implants.

580
00:40:53,480 --> 00:40:55,000
15.

581
00:40:59,560 --> 00:41:01,080
Bone cutter.

582
00:41:07,560 --> 00:41:10,840
Uh, the guide. The drill.

583
00:41:14,960 --> 00:41:17,080
DRILL WHINES

584
00:41:50,000 --> 00:41:51,960
- Everything has gone on well.
- It has?

585
00:41:51,960 --> 00:41:55,480
Everything has gone on well and he is better.

586
00:41:55,480 --> 00:41:58,040
Come. Joshua is here.

587
00:42:03,680 --> 00:42:05,560
SHE SOBS

588

00:42:08,920 --> 00:42:12,480

- Thank you very much.

- Thank you, don't worry.

589

00:42:12,480 --> 00:42:14,120

- Thank you.

- A pleasure.

590

00:42:22,920 --> 00:42:25,560

You can see that the chest is slightly wider

591

00:42:25,560 --> 00:42:27,840

than how it was before. So far, so good.

592

00:42:27,840 --> 00:42:31,160

We have crossed the major part of the bridge,

593

00:42:31,160 --> 00:42:34,840

we still have some more time to go, but at least so far, we're OK.

594

00:42:49,800 --> 00:42:51,760

A little fighter, right enough.

595

00:42:54,880 --> 00:42:57,720

Well, touch wood, wherever there is any,

596

00:42:57,720 --> 00:43:01,320

that everything's going OK at the moment

597

00:43:01,320 --> 00:43:05,000

and hopefully going home today.

598

00:43:05,000 --> 00:43:09,080

To Manchester. How does that sound, Mister?

599

00:43:09,080 --> 00:43:10,680

A little fighter, right enough.

600

00:43:10,680 --> 00:43:14,520

It's not over yet, so we've still got a long, long way to go with him, but...

601

00:43:14,520 --> 00:43:19,080

Yeah, there is a long way, but he's here at the moment, he's doing OK

602

00:43:19,080 --> 00:43:21,880

and just fingers crossed and just hope and pray every day

603

00:43:21,880 --> 00:43:25,840

and just take each day as it comes, cos it is going to be

604

00:43:25,840 --> 00:43:29,960

a long journey, but that's what we're prepared for.

605

00:43:29,960 --> 00:43:32,680

- He's sneaking about.

- Hey, what's up?

606

00:43:32,680 --> 00:43:34,040

Sh.

607

00:43:43,160 --> 00:43:46,400

Joshua is being transferred back to his local hospital

608

00:43:46,400 --> 00:43:48,440

where he will stay to recuperate.

609

00:43:50,360 --> 00:43:55,040

His lungs are adapting well and growing into the space surgeons created in his chest,

610

00:43:55,040 --> 00:43:58,360

but they will have to monitor him closely over the months to come.

611

00:44:20,400 --> 00:44:23,800

Shauna is returning to Great Ormond Street from Middlesbrough.

612

00:44:25,400 --> 00:44:30,280

It's four months since the ethics committee's discussion about Shauna's operation.

613

00:44:30,280 --> 00:44:35,160

Her new windpipe is ready and surgery is scheduled for tomorrow.

614

00:44:37,680 --> 00:44:40,640

So the two primary risks are getting in

615

00:44:40,640 --> 00:44:45,760

and then sorting out how well the graft takes.

616

00:44:45,760 --> 00:44:48,760

That does include a risk to life, as you know.

617

00:44:50,160 --> 00:44:54,280

But I think she's at greater risk not having it done than having it done.

618

00:44:54,280 --> 00:44:56,880

It's really difficult to put numbers on this

619

00:44:56,880 --> 00:44:58,880

when we've only done such a small amount,

620

00:44:58,880 --> 00:45:01,920

but, again, I think you said you understand that before.

621

00:45:01,920 --> 00:45:03,200

I do, yeah.

622

00:45:03,200 --> 00:45:08,320

I know you've thought about this a lot, Shauna, as well. You...

623

00:45:08,320 --> 00:45:12,680

You know that we think we can help you with this, but we're not 100 per cent sure.

624

00:45:12,680 --> 00:45:16,800

We think we can make it better. Last time we spoke you are up for that.

625

00:45:16,800 --> 00:45:20,520

- Yeah.

- Is that still the case?

- Yeah.

626

00:45:22,440 --> 00:45:24,800

OK. I'm sure you've talked about it a lot.

627

00:45:26,600 --> 00:45:30,680

- Are you looking forward to it?

- I think she got a bit scared at the weekend and that,

628

00:45:30,680 --> 00:45:33,360

but we've talked, haven't we?

629

00:45:33,360 --> 00:45:37,640

- You're a brave girl.

- Very brave.

630

00:45:46,880 --> 00:45:48,440

Tomorrow will be dreadful.

631

00:45:50,080 --> 00:45:53,280

It will, it'll be dreadful, but she'll get through it.

632

00:45:53,280 --> 00:45:55,960

I know she will.

633

00:45:55,960 --> 00:45:58,360

She's been through a lot, so I know she'll get through -

634

00:45:58,360 --> 00:46:03,280

well, I'm hoping she'll get through it, but I think she will.

635

00:46:09,240 --> 00:46:11,560

It's the day of Shauna's operation.

636

00:46:11,560 --> 00:46:14,760

A new trachea is being grown in a controlled environment

637

00:46:14,760 --> 00:46:18,520

called a bio reactor, three miles away at the Royal Free Hospital.

638

00:46:19,640 --> 00:46:24,120

It can only survive outside of the laboratory for an hour.

639

00:46:24,120 --> 00:46:28,480

Its arrival at Great Ormond Street must be timed with the removal of her old trachea.

640

00:46:30,200 --> 00:46:33,600

When someone's had as many operations as Shauna's had before,

641

00:46:33,600 --> 00:46:38,520

all the surfaces stick together and it's very...

642

00:46:38,520 --> 00:46:41,000

You have to do a bit of work to separate those surfaces

643

00:46:41,000 --> 00:46:43,360

to make sure you can see all the bits that you need.

644

00:46:43,360 --> 00:46:47,000

The other thing that makes getting into her chest difficult

645

00:46:47,000 --> 00:46:49,800

is that she only has one lung,

646

00:46:49,800 --> 00:46:55,200

so everything... Her heart is shifted over to the side without the lung,

647

00:46:55,200 --> 00:46:58,160

so all the blood vessels which would normally be coming up

648

00:46:58,160 --> 00:47:00,800

in nice, neat little arcs in the middle of the chest

649

00:47:00,800 --> 00:47:03,880

are actually off to one side and curved in the wrong place,

650

00:47:03,880 --> 00:47:07,560

but the point of no return will be when we remove the airway.

651

00:47:07,560 --> 00:47:11,280

We have to put something in its place to get air to go

652

00:47:11,280 --> 00:47:14,400

from the upper part of her body to the lung.

653

00:47:14,400 --> 00:47:17,440

And if, for any reason, there's a disaster at that point,

654

00:47:17,440 --> 00:47:19,000

then we would be in trouble.

655

00:47:48,560 --> 00:47:50,560

We're going to have to move this lung

656

00:47:50,560 --> 00:47:53,200

- to get to the trachea, aren't we?

- Yes, exactly.

657

00:47:53,200 --> 00:47:56,240

- That's the trachea, is it?

- Yes.

658

00:47:56,240 --> 00:48:00,040

- You can see right down to the trachea.

- Feel it. It's like...

- Rock.

659

00:48:00,040 --> 00:48:02,400

If that's the trachea... Jesus Christ.

660

00:48:03,760 --> 00:48:07,000

- Martin, the trachea is like a rock. It's like...
- Is it?

661

00:48:09,320 --> 00:48:12,440

- Well, it feels like the spine.
- Really? My god.

662

00:48:12,440 --> 00:48:14,360

- I thought we were on the spine.
- My god.

663

00:48:17,560 --> 00:48:19,520

We've also got to free up the top

664

00:48:19,520 --> 00:48:23,440

and there's a whole other area stuck down here under the aorta,

665

00:48:23,440 --> 00:48:26,280

so the more of that we can free, maybe we can get a better go at it.

666

00:48:26,280 --> 00:48:29,840

- Scissors, please.
- Hand them back to me.

667

00:48:29,840 --> 00:48:33,760

It's just not very visible what we're doing.

668

00:48:35,200 --> 00:48:36,760

Scissors, please.

669

00:48:43,000 --> 00:48:47,360

The Royal Free Hospital is on stand-by to transfer the new trachea,

670

00:48:47,360 --> 00:48:51,000

but surgery is already running two hours behind.

671

00:48:55,040 --> 00:48:57,880

Oh, hi, Mark. Yeah, it's Martin here.

672

00:48:57,880 --> 00:49:02,960

Martin Elliott's struggling a little bit, well, quite a lot really.

673

00:49:02,960 --> 00:49:05,760

He says it's the most difficult dissection he's ever done,

674

00:49:05,760 --> 00:49:09,480

which is saying something for a man of his experience

675

00:49:09,480 --> 00:49:11,800

and the lumen is absolutely minuscule.

676

00:49:11,800 --> 00:49:14,160

It's a miracle she's been able to breathe.

677

00:49:15,320 --> 00:49:18,160

He thinks we're not going to be sending for the graft

678

00:49:18,160 --> 00:49:20,040

for another hour or so, I'm afraid.

679

00:49:23,720 --> 00:49:26,240

That should be the track for the tracheostomy there.

680

00:49:26,240 --> 00:49:28,600

- Can you feel it?

- Well, I can see it, actually.

- Oh.

681

00:49:30,600 --> 00:49:33,960

After three hours of dissection, Martin has finally exposed

682

00:49:33,960 --> 00:49:37,200

the narrow section of trachea which needs to be replaced.

683

00:49:38,280 --> 00:49:41,600

I think we have to go into bypass next.

684

00:49:41,600 --> 00:49:44,680

The vein is open, Nigel.

685

00:49:44,680 --> 00:49:48,080

Drain. Thank you. There's a little bubble.

686

00:49:49,720 --> 00:49:55,160

A bypass machine will pump oxygenated blood round Shauna's body.

687

00:49:55,160 --> 00:49:58,680

As surgeons remove the trachea, she will not be able to breathe.

688

00:50:00,800 --> 00:50:03,200

Hold those, please.

689

00:50:03,200 --> 00:50:06,040

Somebody hold... Colin? Colin? That's it.

690

00:50:06,040 --> 00:50:09,080

Hold both together, hold both. Really important.

691

00:50:09,080 --> 00:50:11,200

Lift up the suture.

692

00:50:11,200 --> 00:50:13,120

No, keep the suture tense.

693

00:50:14,120 --> 00:50:15,920

Ah. Cramp.

694

00:50:18,560 --> 00:50:21,880

OK. Right, let's get our breath back and have a little stretch

695

00:50:21,880 --> 00:50:23,640

and calm down and we'll be OK.

696

00:50:25,680 --> 00:50:28,920

- Full flow, did you say?

- Four hours into the operation

697

00:50:28,920 --> 00:50:31,760

and Martin has removed most of the old trachea.

698

00:50:34,080 --> 00:50:35,760

I think we need to call them.

699

00:50:37,520 --> 00:50:41,040

OK, we have the call, please, to the Royal Free.

700

00:51:00,160 --> 00:51:03,200

The new trachea can only survive outside the bio reactor

701

00:51:03,200 --> 00:51:06,440

for an hour before it will start to degrade.

702

00:51:42,360 --> 00:51:45,480

This is probably the best bit of her trachea.

703

00:51:45,480 --> 00:51:48,880

You can see that it's really, really, really thick

704

00:51:48,880 --> 00:51:53,560

and there's lots of calcium, and this is the widest part of it.

705

00:51:53,560 --> 00:51:55,960

It should be much thinner-walled

706

00:51:55,960 --> 00:51:58,240

and the hole, the lumen inside,

707

00:51:58,240 --> 00:52:01,120

should be as big as the outside of this.

708

00:52:01,120 --> 00:52:03,840

And this is the best bit. It was very, very stuck.

709

00:52:19,840 --> 00:52:22,560

SIREN WAILS

710

00:52:35,600 --> 00:52:38,920

So now we want to make sure there's nothing sharp

711

00:52:38,920 --> 00:52:43,000

on the back of there and that there's room for the new graft.

712

00:52:43,000 --> 00:52:44,960

If you put your finger behind the aorta

713

00:52:44,960 --> 00:52:48,040

and you will feel masses of dense, fibrous tissue.

714

00:52:49,800 --> 00:52:51,800

We have to make sure there's enough space.

715

00:52:51,800 --> 00:52:54,360

What we can't have is the thing kinked by rigid tissue.

716

00:53:06,720 --> 00:53:09,400

'Arriving at destination on right.'

717

00:53:13,720 --> 00:53:18,000

It takes half an hour for the trachea to arrive at Great Ormond Street.

718

00:53:18,000 --> 00:53:22,480

They now have another 30 minutes to transplant it in an optimum condition.

719

00:53:44,680 --> 00:53:47,520

We are ready for the graft.

720

00:53:47,520 --> 00:53:51,400

This is the top. Yeah?

721

00:53:58,840 --> 00:54:00,680

It's just beautiful.

722

00:54:14,080 --> 00:54:19,120

The lungs are coming up beautifully and there's no air leak at all.

723

00:54:19,120 --> 00:54:22,920

Can you see inside? There's a join there we've made with stitches.

724

00:54:25,680 --> 00:54:28,840

After eight hours of surgery, Shauna is off bypass

725

00:54:28,840 --> 00:54:31,400

and breathing through her new trachea.

726

00:54:33,560 --> 00:54:36,120

Can I just say while everybody's having a quiet moment,

727

00:54:36,120 --> 00:54:39,280

thank you very much, everybody - you've worked your butts off today

728

00:54:39,280 --> 00:54:42,440

and it's been really appreciated. Really, really nice. Thank you.

729

00:55:02,720 --> 00:55:04,440

Oh, you're brave, Shauna.

730

00:55:10,360 --> 00:55:16,200

- Thank you.

- Thank you very much.

- Take care. Sleep well.

- Thank you.

731

00:55:32,360 --> 00:55:36,120

Two weeks after her surgery, and Shauna is well enough

732

00:55:36,120 --> 00:55:39,640
to go back to Leeds hospital where she will begin her rehabilitation.

733
00:55:39,640 --> 00:55:41,720
More emergency stuff.

734
00:55:41,720 --> 00:55:45,360
Can you take care of that, Shauna? For the way down, yeah?

735
00:55:45,360 --> 00:55:47,600
OK.

736
00:55:47,600 --> 00:55:50,040
Let's rock and roll it, OK?

737
00:55:54,240 --> 00:55:56,600
Bye, Richie.

738
00:55:58,560 --> 00:56:01,800
Whilst her transplant has so far been successful,

739
00:56:01,800 --> 00:56:04,360
the next few weeks will be critical.

740
00:56:37,280 --> 00:56:39,040
It's just a tragedy.

741
00:56:39,040 --> 00:56:42,960
I've feel so sorry for Shauna's family and for her.

742
00:56:42,960 --> 00:56:45,120
Do you regret doing it?

743
00:56:46,160 --> 00:56:47,880
Um, no, I don't regret doing it

744
00:56:47,880 --> 00:56:51,360
because we knew from what was happening to Shauna beforehand

745
00:56:51,360 --> 00:56:54,480
that she'd reached the end of conventional therapy.

746
00:56:56,040 --> 00:57:00,280
You ask yourself this sort of thing all the time if something bad happens,

747

00:57:00,280 --> 00:57:04,040
but if you don't try for that individual patient,

748
00:57:04,040 --> 00:57:06,720
then you can make no progress.

749
00:57:06,720 --> 00:57:12,200
Every patient who survives is standing on the shoulders of people who didn't.

750
00:57:12,200 --> 00:57:16,760
Every operation we do learns from the experience of the previous one.

751
00:57:16,760 --> 00:57:19,240
The more you do, the better you get.

752
00:57:20,320 --> 00:57:24,720
Each of those sounds like a soundbite,

753
00:57:24,720 --> 00:57:27,320
but they are all true.

754
00:57:27,320 --> 00:57:31,040
The fundamental core principle of this is

755
00:57:31,040 --> 00:57:35,000
is this the right general strategy to develop?

756
00:57:35,000 --> 00:57:39,920
I don't think we have any doubts of that. We've talked about it a lot.

757
00:57:39,920 --> 00:57:43,880
Should we abandon the sort of therapy or carry on?

758
00:57:43,880 --> 00:57:50,680
And we are, as a group, convinced that we should carry on.

759
00:58:22,880 --> 00:58:25,920
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