Great Ormond Street Children's Hospital has helped develop cures for many conditions which just a few years ago were untreatable. But for medical science to move forward, doctors must be able to experiment. We've done a very small number of patients with this condition in England. If you sign the consent form, you're basically signing a contract with uncertainty. Experimental surgery on children raises difficult ethical dilemmas. Whenever you have any patient, it's trying to make the right decision and when they are so severe, possibly the right decision is not to operate on them. For children with conditions which have no known cure, these experiments are their only hope. This is basically our only option to give him a better life, otherwise... he'll die. Surgeons are constantly forced to question how far
they should push the boundaries in the hope of finding a cure.

Sometimes we've got the technology, we can do all kinds of things, but we have to also ask the question, is it the right thing to do?

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

15-year-old Shauna has spent her entire life in and out of hospital. She was born with one lung, a major heart defect and her windpipe is slowly closing.

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

Do you want to ring the doorbell?

Recently, her ability to breathe has deteriorated and she is at risk of suffocation.

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

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

I nearly lost her that day. It took them seven minutes to get her back.
If they don't do anything pretty quick, I won't have her much longer. They've told me that.

State-of-the-art equipment, huh? You've got a telly in here as well. Shauna is at Great Ormond Street to see if she is eligible for experimental surgery that has only been performed on one other child. Surgeons want to offer her a donor trachea which will be modified with Shauna's own stem cells. If they go ahead, she will be given a new windpipe made of her own DNA.

We've only done one tracheal transplant before and we learned a lot of things in a very short period of time and there are no long-term data. The balance between is this appropriate or ethical, right or wrong, is a scenario in which there are no correct answers. There's just best judgement and so that's what we'll try to achieve
between us all and get a lot of input into getting to that decision.

Colin Wallace is a respiratory consultant at Great Ormond Street.

Hi.

He's part of the team weighing up the risks of the transplant and her current quality of life.

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

- We haven't met before, have we?
- No.

If I was to ask you for three things that you would like to have better after an operation on your trachea, what would they be?

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

Name one of them - what would you like?

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

What do you miss? You like to do?

- You go with the carers, but you can't do it.
- Swimming.
You'd like to try swimming? And you go to school? Normal school?

Do you have to have someone with you?

Yes.

What's that like?

All right.

Hm?

All right.

It's OK, yes? And at home?

She has night carers.

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

so that I can have some rest.

Yes. And you have a friend at school?

What's your best friend's name?

Courtney.

What did she think about your tracheostomy?

She's all right with it.

She's used to it now, is she?

Yes.

And what about other things you'd like to do
that you can't do at the moment?

73
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.

75
00:04:47,120 --> 00:04:52,000
- Yes.
- You think so? Would that be quite an adventure, hey?

76
00:04:52,000 --> 00:04:56,440
What would you need to know will happen

77
00:04:56,440 --> 00:04:59,120
to be able to go ahead and say yes, we'll have the surgery?

78
00:05:01,400 --> 00:05:02,640
Well, it's stupid really,

79
00:05:02,640 --> 00:05:05,120
cos I'd want to know if it was going to be a success.

80
00:05:05,120 --> 00:05:09,080
- But you can't give me that guarantee.
- Yes.

81
00:05:09,080 --> 00:05:11,840
So, because her life is good at the moment,

82
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.

86
00:05:20,960 --> 00:05:25,320
It shows there's a vulnerability here on the narrowing of the trachea

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

Yes.

Shauna is in charge. You in charge? Yay!

What's quite interesting is that mother does perceive
the current quality of life as being good.

In other words, this is not a situation where we've got
nothing to lose by going ahead - they've got quite a bit to lose

and this is going to make for a difficult decision.

She's got a reasonable quality of life, she's got her good friend,
she clearly has a sense of humour and enjoying herself -

it makes it harder.

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

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

112
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.

114
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
that's supporting it, but inside the trachea, it's very narrow.

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

It's lying perilously close to very big vessels

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

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

Encouragingly,

the airways from the trachea onwards are of good calibre

and nicely open.

But my feeling is that on these scans,

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

This lung could cope.

As tracheal transplants in children are so new,

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

before offering surgery.

It is made up of professionals from a variety of backgrounds,
from both inside and outside the hospital.

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

Frankly, we are so focused on trying to make her better that we need a more cool and detached intellectual discussion.

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

We took a donor trachea and that trachea was then washed until all the cells were removed from it,

so detergent enzymatic washes.

So you're left with what is essentially a scaffold of largely collagen and just a few proteins,

but no active... immunologically active cells.

That graft was marinaded with stem cells and then locally we injected a drug called TGF beta,

which is supposed to trigger it to become cartilage.

So after it's been marinaded, the graft returned in a motorbike.
to Great Ormond Street and we were able to stitch the trachea into Kieron.

So what we want to do for Shauna is to use the same basic principles of allowing a skeleton of tissue to be populated by the child's own stem cells, so that there's no rejection, and to get more rapid cellularisation of the child than we had time for in Kieron.

I'd just like to know, if you don't do the procedure, what is likely to be the manner of her death? What happens as the winter emerges is that the airway, the trachea is very like your nose, so as you imagine your nose blocking up when you have a cold, if you've ever...just remember breathing through a snorkel for the first time, the sense of not being able to breathe is...

One of the greatest fears that you can have, so I can't imagine that it would be a pleasant demise.
But she's going to die anyway, even if you do the procedure -

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

Um...

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

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

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

Or at least we prolong her life

And her quality of life for as long as possible.

In terms of the timing of the decision-making,

would there be any advantages in deferring?

Would it give better outcomes or whatever?

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

The section here is only 2mm across.

That's the tip of a Biro.

At that point, you have to really consider how much
swelling of the mucosa you can tolerate before it becomes a critical narrowing.

You haven't mentioned palliative care as an option. Is that because you think it's inappropriate?

No, I think if I thought the treatment was really horrific, and we hadn't got the experience that we have had, I wouldn't have any hesitation in offering that.

I don't offer palliative care for complex patients. So it's not really just a choice between a horrible death and the chance of improved quality of life?

There is a third option, which is a less horrible death?

Yes, that's true. Palliative care may still become necessary, but at the moment I'm not sure that the technical solution does not indeed trump the palliative care.
But only for the willing participant in uncertainty.

Do we have any further questions?

The committee debated for another hour and found no ethical objections to Shauna’s surgery.

Shauna will return to Middlesbrough and may have to wait up to four months for the new trachea to be grown.

Three-month-old Muhammed has just arrived on the cardiac intensive care unit. At birth, he was diagnosed with Jeune’s Syndrome, a very rare condition with only 125 recorded cases worldwide.

Muhammed’s rib cage doesn’t grow. It is trapping his lungs and eventually, he will be unable to produce enough oxygen to stay alive.

Can we drop these lights now?
The width of the abdomen is normal width for a child and if you just imagine where the soft tissue ends on the right with the vague, grey things on either side, that would be the normal width.

So that's baby size.

If you come up towards the chest, the chest narrows right in and then comes out again in the armpits. This is characteristic of Jeune's Syndrome.

So this chest here is squeezing in and the heart in the middle and the lungs are actually squashed, so the child's chest is going to stay the same size.

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

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

After searching on the Internet, Muhammed's father read about an experimental trial at Great Ormond Street,
where they expand the rib cage in the hope that the lungs will grow.

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

and has been performed on just six children before now.

- Hello, hi.
- Hi.
- My name is Martin Elliott.

Hi, very nice to meet you.

- Nice to meet you.

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

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

of doing this using Neil's hands.

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

and reconnect them...

with plates and screws,

so effectively,

we're going from four ribs, to two ribs, which are longer.
We would cut the third one in the middle,

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

so that we could make them longer

but leave the third and the bottom one loose.

So, it's just stretching it out.

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

and make his chest wall even bigger.

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

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

All got back home.

One of them has sadly died later

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

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

which is so uncertain.

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

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

What we can do as parents.

I think that's the main thing.

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

And...

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

and make sure he's all right.

It is not compulsory to have treatment for this

because no-one knows what the future is.

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

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

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

The option is to accept that death is inevitable.

I know the Birmingham team have even put that in a more explicit way
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.
- We think there, probably, don't we?  
- Yep.

Down to eight.

There won't be much room, will there?

Gosh, it's tight.

It's amazing they can breathe at all when you put so much force on here.

Diathermy, please.

So, we've just, having cut those ribs,

and now we've brought them forwards, so this is one rib up

and one rib down, joined together, so if I let it go, it forms an arch.

So it's lifted the whole chest wall by a good two centimetres.

Once Neil Bulstrode can see the expanded ribs,

he can then shape the titanium plates which will hold them together.

That's good. That's going to be great. OK, the clamps, please.

That one, yep. And we need a syringe of saline.
The drill didn't go all the way through the...

This is the eight, please.

DRILL WHIRRRS

Eight, please.

Great.

Just check for any...

The titanium plate has screwed in beautifully.

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

they've now been rejoined to lengthen the rib.

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

So we get them to stage two and then,

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

through a separate incision and see how we get on.

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

I think that's all right.
If we just stitch the other stuff up, it'll be OK, won't it?

Yeah, yeah.

It has taken one and a half hours to break and expand the left side of Muhammed's rib cage.

They now have to do the same to the right.

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

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

so we basically want to get it right first time.

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

Anne, the anaesthetist, said that the ventilation had got enormously better instantly.

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

it should be greatly improved.

After three hours of surgery,

Muhammed is taken back to intensive care.
He will remain on a ventilator and sedated for the next few days.

His lungs will be monitored round-the-clock to see if they adapt to his expanded rib cage.

The expansion on both sides went according to plan and immediately the anaesthetist said there was an improvement in the ability to ventilate Muhammed,

- so we were really pleased with that.
- Yeah, yeah.

- He's doing really well.
- That's a really good news, yeah.

- I was really concerned.
- Of course.

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

Do you have any other questions?

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

- He was coughing a lot.
- Right.
- Would he still have the same issues?
We’ll have to see exactly how that goes.

Sometimes we can put another little tube in through his nose which goes into the stomach and, if necessary, we can feed him through there temporarily.

- You can tell the difference with the chest already.
- Chest, yeah.

You can already see the increase in the size.

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

Three days after surgery, Muhammad’s lungs are struggling to expand in his new rib cage.

The ventilation’s still not perfect, you know, so there’s still some episodes where the SATs fall and some episodes where it’s a bit hard to get air into the lungs, and also, I guess, he’s not quite moving as much air yet as we thought, but it’s only a few days since he had a big operation and we broke quite a lot of ribs,
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.

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

What we have to have is an understanding of uncertainty.

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

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

and his breathing is becoming increasingly difficult.

We've probably got to the end of the road

as far as what we can do for him.

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

another operation to enlarge his chest in any other way

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

Doctors in Birmingham have told Muhammed's father

they want to take him off life support.

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

and a conference call has been arranged.
It's very difficult to know how to help the family through this. There isn't a way. It's just painful.

Um, hi, Professor. Hello.

Yeah.

Well, um, what I said to you was I didn't know whether the lungs would grow at all and, to be certain that there's going to be some improvement, you need a long period of time, but if there's been deterioration in between times I think that really gives us the answer that there just simply isn't enough lung for Muhammed to be able to survive on his own.

OK.

OK.

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

OK, well...

We all send you our best wishes from here.

- Thank you.
- Thank you.

- Bye.
- Thanks a lot. Cheers. Bye.

HE SIGHS

OK.

Would we do it again?

For the time being, if we accumulate enough patients in this category to say, "Look, this is never going to work."

"This category of patients are inappropriate for this therapy because of X or Y",

then clearly that would be relevant, but I don't think we're there yet.

We're still trying to find out what X and Y are that would stop you doing something.
A few months later and another case of Jeune's syndrome has been transferred from Stepping Hill Hospital to Great Ormond Street.

Joshua Burns Adair, he's five months old now and has come down for another expansion tomorrow.

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

been up to a maximum of 90% oxygen and he was in 45% when we picked him up.

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

Joshua's condition is critical and despite the dangers, his parents are eager for the chest expansion.

Surgery is scheduled for tomorrow morning.

After doing everything on the internet, I brought it up with the doctors and I pushed them to look into it and it was via, basically, our consultant finding out that there was actually a surgeon down here
that can do this operation.

- We decided that we wanted to go down every avenue, didn't we?
- Yeah.

To see that we knew if anything did happen to Joshua,

we'd done everything we could.

So this is basically our only option to give him a better life,

otherwise...he'll die.

Whilst Joshua is being transferred from the ambulance ventilator
to one on intensive care, his heart stops beating.

It takes two minutes to revive him.

The operation is cancelled

and his parents are left waiting for the surgery to be rescheduled.

The way they've been speaking for the last month when we had telephone conversations,

their hopes are very high we can fix him, even though

they have been spoken to and they know that this is experimental.
But Joshua is different now than he was two days ago and I think if he doesn't get back to his transfer settings then the surgery is not an option and we have to go down that avenue. The issue for us and the reason that we're isolated is that the CO2 is all over the place and ultimately that's not good for the lungs and it's not going to be good for multiple procedures so I would think it would be daft to operate in the next day. Yeah, I mean, I would just say do we, after we've got through this next 48 hours, see if we can optimise him as much as we can and then we know where we are. We should then reappraise whether we should actually offer surgery or not. I think we just need to wait until we've got that. When he's ready for a general anaesthetic.
I'm not sure what we can do at this stage.

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

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

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

Yeah.

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

- Being left to his own, sort of, agenda, as such?
  - Yes.
  - Until they...

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

You know, whenever you have any patient,

it's trying to make the right decision

and when they are so severe...

Possibly the right decision is not to operate on them

and I think if Joshua stays in his current state

that that'll be our answer.
However, we're all optimistic people that want to do things to try and help, but sometimes we have to step back and admit that maybe we cannot help them. Always wore a red cloak with a warm hood and so she was called Little Red Riding Hood. One day she decided to visit her granny who lived some way from the woodcutter's cottage. She took a basket with a cake her mother had baked and set off. Now, the last thing her mother had said to Little Red Riding Hood was, "Don't leave the path and don't talk to any strangers." I'm afraid Little Red Riding Hood was not really listening. 48 hours later and Joshua's oxygen levels have improved. I guess the key discussion for us is he's improved, has he improved to the point we can do surgery?
Is this our window for surgery and if we wait are going to miss that?

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

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

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

because things may not get much better.

I completely agree that this may be the window

that we are actually getting

for treating Joshua in the right direction,

but if anything can be done, probably,

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

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

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

That is a very real possibility as well.

You know, we should continue to also say

that if they felt that they didn't want to go down this route
given the risks now that we would fully support them in that decision as well.

They have to know that he has about a one in two chance of coming through.

You're semi-detached from this, Ruth.

Well, I am.

- Are you disturbed by it?
- Yes, to be honest.

I've just come into this and don't really know the case,

but I have to say I am kind of taken aback about going forward

for such dramatic surgery with a risk of 50%.

That's... And I am detached. You know, I don't know the child.

It's a good point. On the other side of the coin is, sort of,

the alternative is 100% chance of death within a year probably.

I'd be very surprised if this child was alive in six months, even,

if he didn't have surgery.

And so that's the... The risk is in that context.

That, sort of, to me... On the one hand, is this whole thing crazy?

This is right at the edge of what I feel we're fully comfortable with doing.
Really right at the edge of it and...

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

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

but as long as the parents are absolutely clear

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

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

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

and so that's quite a lot more encouraging

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

Now, that doesn't by any means mean

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

I get the feeling we're at the level,

the best we could have hoped for

So I think we've got to a point

where if we're going to do it,
probably now is the best time we can.

There are some risks that we do not know.

As I said, the experience on the surgery's quite dangerous.

The known risks are there are chances which are very high

that he will not come through, or he will not make it after surgery alive.

Other risks, which we know definitely,

are Joshua will need multiple surgeries

and at any stage he may fail to cope with all these interventions. We need to be aware of that.

Our own experience, we have lost two children.

When we say 50%, what we're saying is it's very high, you know?

It means if you took two children into the operating theatre,

only one of the children would come back out,

but the other thing that people felt we should also just discuss

is if things sadly go wrong in theatre,
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,
you should be strong enough to carry on.

There's no backing out.

My brave little soldier. Aren't you?

You know, to your mother, you're everything in the world.

You show them. Yeah? You show 'em.

This is Joshua Burns Adair.

Consented for lateral chest-expansion on both sides with metallic implants.

Bone cutter.

Uh, the guide. The drill.

DRILL WHINES

- Everything has gone on well.
- It has?

Everything has gone on well and he is better.

Come. Joshua is here.

SHE SOBS
- Thank you very much.
- Thank you, don't worry.

- Thank you.
- A pleasure.

You can see that the chest is slightly wider than how it was before. So far, so good.

We have crossed the major part of the bridge,

we still have some more time to go, but at least so far, we're OK.

A little fighter, right enough.

Well, touch wood, wherever there is any,

that everything's going OK at the moment

and hopefully going home today.

To Manchester. How does that sound, Mister?

A little fighter, right enough.

It's not over yet, so we've still got a long, long way to go with him, but...

Yeah, there is a long way, but he's here at the moment, he's doing OK
and just fingers crossed and just hope and pray every day

and just take each day as it comes, cos it is going to be a long journey, but that's what we're prepared for.

- He's sneaking about.
- Hey, what's up?

Joshua is being transferred back to his local hospital where he will stay to recuperate.

His lungs are adapting well and growing into the space surgeons created in his chest,

but they will have to monitor him closely over the months to come.

Shauna is returning to Great Ormond Street from Middlesbrough.

It's four months since the ethics committee's discussion about Shauna's operation.

Her new windpipe is ready and surgery is scheduled for tomorrow.

So the two primary risks are getting in and then sorting out how well the graft takes.

That does include a risk to life, as you know.
But I think she's at greater risk not having it done than having it done.

It's really difficult to put numbers on this when we've only done such a small amount,

but, again, I think you said you understand that before.

I do, yeah.

I know you've thought about this a lot, Shauna, as well. You...

You know that we think we can help you with this, but we're not 100 per cent sure.

We think we can make it better. Last time we spoke you are up for that.

Yeah.

- Is that still the case?
- Yeah.

OK. I'm sure you've talked about it a lot.

- Are you looking forward to it?
- I think she got a bit scared at the weekend and that,

but we've talked, haven't we?

- You're a brave girl.
- Very brave.

Tomorrow will be dreadful.
It will, it'll be dreadful, but she'll get through it.

I know she will.

She's been through a lot, so I know she'll get through -

well, I'm hoping she'll get through it, but I think she will.

It's the day of Shauna's operation.

A new trachea is being grown in a controlled environment called a bio reactor, three miles away at the Royal Free Hospital.

It can only survive outside of the laboratory for an hour.

Its arrival at Great Ormond Street must be timed with the removal of her old trachea.

When someone's had as many operations as Shauna's had before,

all the surfaces stick together and it's very...

You have to do a bit of work to separate those surfaces to make sure you can see all the bits that you need.

The other thing that makes getting into her chest difficult is that she only has one lung,
so everything... Her heart is shifted over to the side without the lung,

so all the blood vessels which would normally be coming up

in nice, neat little arcs in the middle of the chest

are actually off to one side and curved in the wrong place,

but the point of no return will be when we remove the airway.

We have to put something in its place to get air to go

from the upper part of her body to the lung.

And if, for any reason, there's a disaster at that point,

then we would be in trouble.

We're going to have to move this lung

- to get to the trachea, aren't we?
- Yes, exactly.

- That's the trachea, is it?
- Yes.

- You can see right down to the trachea.
- Feel it. It's like...
- Rock.

If that's the trachea... Jesus Christ.
- Martin, the trachea is like a rock. It's like...
- Is it?

- Well, it feels like the spine.
- Really? My god.

- I thought we were on the spine.
- My god.

We've also got to free up the top

and there's a whole other area stuck down here under the aorta,

so the more of that we can free, maybe we can get a better go at it.

- Scissors, please.
- Hand them back to me.

It's just not very visible what we're doing.

Scissors, please.

The Royal Free Hospital is on stand-by to transfer the new trachea,

but surgery is already running two hours behind.

Oh, hi, Mark. Yeah, it's Martin here.

Martin Elliott’s struggling a little bit, well, quite a lot really.

He says it’s the most difficult dissection he’s ever done,
which is saying something for a man of his experience

and the lumen is absolutely minuscule.

It's a miracle she's been able to breathe.

He thinks we're not going to be sending for the graft

for another hour or so, I'm afraid.

That should be the track for the tracheostomy there.

- Can you feel it?
- Well, I can see it, actually.
- Oh.

After three hours of dissection, Martin has finally exposed

the narrow section of trachea which needs to be replaced.

I think we have to go into bypass next.

The vein is open, Nigel.

Drain. Thank you. There's a little bubble.

A bypass machine will pump oxygenated blood round Shauna's body.

As surgeons remove the trachea, she will not be able to breathe.
Hold those, please.

Somebody hold... Colin? Colin? That's it.

Hold both together, hold both. Really important.

Lift up the suture.

No, keep the suture tense.

Ah. Cramp.

OK. Right, let's get our breath back and have a little stretch and calm down and we'll be OK.

- Full flow, did you say?
- Four hours into the operation

and Martin has removed most of the old trachea.

I think we need to call them.

OK, we have the call, please, to the Royal Free.

The new trachea can only survive outside the bio reactor for an hour before it will start to degrade.

This is probably the best bit of her trachea.
You can see that it's really, really, really thick and there's lots of calcium, and this is the widest part of it. It should be much thinner-walled and the hole, the lumen inside, should be as big as the outside of this. And this is the best bit. It was very, very stuck.

So now we want to make sure there's nothing sharp on the back of there and that there's room for the new graft. If you put your finger behind the aorta and you will feel masses of dense, fibrous tissue. We have to make sure there's enough space. What we can't have is the thing kinked by rigid tissue. 'Arriving at destination on right.'

It takes half an hour for the trachea to arrive at Great Ormond Street.
They now have another 30 minutes to transplant it in an optimum condition.

We are ready for the graft.

This is the top. Yeah?

It's just beautiful.

The lungs are coming up beautifully and there's no air leak at all.

Can you see inside? There's a join there we've made with stitches.

After eight hours of surgery, Shauna is off bypass and breathing through her new trachea.

Can I just say while everybody's having a quiet moment,

thank you very much, everybody - you've worked your butts off today and it's been really appreciated. Really, really nice. Thank you.

Oh, you're brave, Shauna.

Two weeks after her surgery, and Shauna is well enough
to go back to Leeds hospital where she will begin her rehabilitation.

More emergency stuff.

Can you take care of that, Shauna? For the way down, yeah?

OK.

Let's rock and roll it, OK?

Bye, Richie.

Whilst her transplant has so far been successful,

the next few weeks will be critical.

It's just a tragedy.

I've feel so sorry for Shauna's family and for her.

Do you regret doing it?

Um, no, I don't regret doing it because we knew from what was happening to Shauna beforehand that she'd reached the end of conventional therapy.

You ask yourself this sort of thing all the time if something bad happens,
but if you don't try for that individual patient,

then you can make no progress.

Every patient who survives is standing on the shoulders of people who didn't.

Every operation we do learns from the experience of the previous one.

The more you do, the better you get.

Each of those sounds like a soundbite,

but they are all true.

The fundamental core principle of this is

is this the right general strategy to develop?

I don't think we have any doubts of that. We've talked about it a lot.

Should we abandon the sort of therapy or carry on?

And we are, as a group, convinced that we should carry on.