

Broken Hearts

Cold Open

Hannah: Inside SickKids, Bravery Beads are as ubiquitous as masks and scrubs. Some of the chunky necklaces dangle from wheelchairs and IV poles. Others hang from patients' necks, a brightly coloured reminder of how far they've come—and how much they've endured. Because Bravery Beads aren't given. They're earned.

A bead is awarded for each procedure, pinprick, and test, from bloodwork to chemotherapy. Collectively, they tell a story.

For three-year-old Annabeth, it's a long one. She has at least nine necklaces, each thick with beads of many shapes and colours. That's because Annabeth was born with hypoplastic left heart syndrome, a serious congenital defect.

Here's how her mom Raven explains it:

Raven: A normal heart would have the right and left atrium and right and left ventricle. So, her left ventricle is basically nonexistent. The little valve between her atrium and ventricle was completely closed and her left atrium was like a third, maybe, maybe a quarter of how big it was supposed to be.

Hannah: Annabeth had her first operation at four days old. Since then, she's had many more, from valve replacements to the implantation of a permanent pacemaker. But all the operations, procedures, and medications, they're stopgap measures. What Annabeth really needs is a new heart.

[MUSIC]

Hannah: Welcome to SickKids VS, where we take you to the frontlines in the fight for child health. I'm Hannah Bank. And this is SickKids VS Broken Hearts.

ACT ONE

Hannah: In April of 2019, three-year-old Annabeth is in heart failure, living inside the winding Cardiac Critical Care Unit at SickKids. A breathing mask hugs her face as she plays with her doll.

Annabeth was listed for a donor heart a few months ago. Since then, Raven and her husband Cameron have been anxiously waiting.

Raven: You're always wondering, is this going to be the call?

Hannah: Annabeth is one of roughly fifty kids waitlisted for a heart in Canada each year. Of those, 30-35 will get one. Others will continue waiting, some for years. And many will die in the process.

But why do kids have to wait so long?

Dr. Osami Honjo: We used to have a more donor availability in the past. And this is a worldwide trend that the number of donor or donation is declining over time.

Hannah Bank: That's Dr. Osami Honjo. He's a researcher and heart surgeon at SickKids. This is a problem he thinks about. A lot.

Dr. Osami Honjo: And I think one good thing is world is it getting safer. Now everybody gets a seat belt and so I think there are more awareness about you know infants Sudden Death Syndrome, for example. But we don't know the exact reason.

Hannah Bank: To expand the organ donor pool, [the Canadian Medical Association approved donations after circulatory death, or DCD, in 2006](#). Before, doctors could only harvest organs *after* brain death. Now, they can harvest them once the heart's stopped beating, too. [That's led to a huge increase in available organs—except hearts](#). Because DCD hearts tend to be starved of oxygen and blood, which makes them a lot less viable.

So, the availability of donor hearts is one problem. The other is geography.

Say SickKids gets a call from Trillium Gift of Life, an organ donation network. They've got a potential donor heart in Vancouver—a four-and-a-half-hour flight from Toronto. SickKids confirms the donor heart's a good match. Next, two surgical teams form—one in each city. They plan and coordinate. Because, for this to work, it must be as precisely timed and executed as a military operation.

While the Vancouver team harvests the heart, the Toronto team preps the patient for transplant.

Dr. Honjo: And when everybody is ready, this surgeon on site use a medication to stop the heart and then explant from the donor body. And then put in the bucket and then they rush back to the hospital here.

Hannah: That's when the clock starts ticking. Because even while the heart is immersed in a cooling solution, sealed in plastic and sitting on ice, it's degrading. This period, when the heart is deprived of blood flow, is known as ischemic time.

Dr. Honjo: And that has to be roughly within six hours. So that's the biggest limitation. And if you exceed six hours, then the heart tend to be very weak or muscle get permanent damage so that never pump properly.

Hannah: There's nothing Osami can do about the shortage of donor hearts. But he thinks there *is* something he can do about ischemic time, which would even improve DCD hearts. It's a way to keep the heart beating *outside* the body by feeding it warm, oxygenated blood. Theoretically, it could reduce ischemic time to almost zero.

The technical term is ex vivo perfusion. But Osami likes to call it the Heart in a Box.

ACT TWO

Hannah: Osami is a clinician scientist. He sees patients *and* works in the lab. He looks for practical solutions to everyday problems like donor heart availability. Because he cares about his patients. And he's seen some of them die waiting.

Raven knows how much Osami cares because he's Annabeth's surgeon.

Raven: He is always more than willing to come and talk to us at the bedside. If he had any questions, he would come in between his surgeries. As soon as you tell him, oh, her echo's been done. He'll like run to the closest computer and look at the results and he'll point out all of the different things that have changed between her last echo and this one. And he will just come and visit all the time.

Hannah: So, when Osami starts working on the Heart in a Box, the problem isn't distant or abstract. It's something he's seen firsthand.

Fortunately, there's a precedent for such a device. In fact, there's already a commercially available ex vivo perfusion machine. It's called [the Organ Care System](#). It's a sleek, high-tech device, which looks like something designed by Steve Jobs and a mad scientist. On its website, you can see it in action. A digital screen on top, tubes pumping blood, and a heart cocooned in sterile plastic, beating with a steady rhythm. It's incredible.

So, why can't Osami just use that? Why are they starting from scratch instead?

Dr. Honjo: The Organ Care System is essentially purely designed for the adult heart. It's, it's huge.

Hannah: The average adult heart is about the size of a bell pepper. The average three-year-old heart is closer to a plum. There are other differences between adult and child hearts too.

Marlee Parker: Paediatric hearts, structurally, from their cardiac cells, differ from adult cells and so you can't even really use the same strategies most times.

Hannah: That's Marlee Parker. She's a perfusionist, which is someone who operates the machinery during surgery that keeps the heart and lungs functioning while surgeons operate. Which comes in handy, because the bedrock of the Heart in the Box is the ECMO, which stands for Extracorporeal Membrane Oxygenation.

When a heart is stopped during surgery, the ECMO feeds it warm, oxygenated blood to keep it functioning. But it's not built to keep a heart alive *outside* the body. To get it to do that, Osami's team needs to retool it, to hack it. They add tubing to create a circuit for blood flow. And an off-brand Tupperware box to hold the heart. They tweak the volume, pressure, and flow speed. And begin testing it.

Dr. Honjo: We obviously struggle to get this perfusion going. I'll say first five heart didn't really move on the device and we were just wondering *why*.

Hannah: It's frustrating. But the team persists. They make more tweaks, adjusting the flow pressure and blood volume. Each time it gets a bit better.

Around the sixth test, they connect the heart and activate the machine. Then, they watch the motionless heart in a box, waiting. They're in a lab, but it's built to look and feel exactly like an operating

room. Same bright lights, same mechanical beeps, same stainless-steel operating table. All of which seem to add to the tension.

Suddenly, the heart twitches awake.

Marlee Parker: It's sluggish and it's trying to wake up and then it then it gets going it's, it's pretty amazing.

Hannah: It's a thrilling moment. Some cheer. Others get emotional.

Marlee Parker: I teared up a little bit because I was the first time I'd ever seen a heart beat on its own outside of the natural body.

Hannah: The more tests they run, the more they improve. Now, they can keep a heart beating for *hours* outside the body. But they haven't successfully transplanted one of the hearts. Nor are they quite ready for human trials.

Which means, for now, kids like Annabeth have to keep waiting.

MID-ROLL COMMERCIAL

Andrew: Children aren't small adults. They require specialized care. And paediatric doctors like Osami are often forced to play catch-up, redesigning existing tools so they work for kids. All that takes time—and money. Some of that money comes from grants and governments. But a lot of it comes from people like you. So, if you want fund work like Osami's Heart in a Box, donate to SickKids at sickkidsfoundation.com/podcast.

ACT THREE

Hannah: It's April 2019. Three-year-old Annabeth is in heart failure. Another month or two, and she'll probably be too sick to be a transplant candidate. Raven, Annabeth's mom, waits anxiously, her phone nearby.

Then the call comes.

Raven: I thought they were joking at first. I was like, "No, you're. You told us we were going to be on the waiting list for over a year." They were like, "No, she needs the heart and she's going to get it."

Hannah: But the surrealism of the moment soon yields to different emotions—hope, relief, but also sadness.

Raven: You're waiting for someone else's kid to die. So as thankful as we can be. You do. You spend however long waiting for that call to happen, but you're also knowing that somebody else had to say yes.

Hannah: Dr. Osami Honjo performs the surgery. It lasts a gruelling 12 hours. But it works. Annabeth has a new heart.

Over the next few months, Annabeth suffers from multiple transplant complications, on top of all the leftover damage from her original heart. She's had medication to help her blood pressure, dialysis to relieve her kidneys, and a tracheotomy to help her breathe.

In November, six months after Annabeth's transplant, I meet Raven in the Cardiac Critical Care Unit at SickKids. I ask her how Annabeth's heart is.

Raven: It's great. It's the best part of her. Other than her brain, her heart and her brain are the great, great parts of her. And her surgeon is extremely happy.

Hannah: Osami saved Annabeth's life. But that was only possible because a donor heart was available. Not every kid is so lucky.

With the Heart in a Box, Osami hopes to change that. He knows there's still a long way to go. There's still more research to do, still more tests to run.

But he's getting closer. And he's not giving up.

EXTRO

Hannah: From SickKids Foundation, this is SickKids VS. Thanks for listening.

If you want to support work like this, visit sickkidsfoundation.com/podcast to donate. And if you like this podcast, please subscribe and rate us on Apple Podcasts, Spotify, or wherever you listen to SickKids VS.

SickKids VS is produced by me, Hannah Bank, Kate Daley, Colin J. Fleming, and Gillian Savigny. This episode was written by Colin J. Fleming.

Sound design and editing by Sndwrx.

Production support by Ayesha Barmania.

For photos, transcripts, sources, show notes, and lists of donors as well as researchers and clinicians who helped make this breakthrough possible, visit sickkidsfoundation.com/podcast.

Dr. Osami Honjo is the Watson Family Chair in Cardiovascular Sciences.

Dr. Junko Kobayashi, who worked with Osami on this project, is the inaugural Dr. William Mustard Fellow in Cardiovascular Surgery Research.