

Biliary Atresia

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1. Biliary Atresia - What is Biliary Atresia, the causes, and treatment_

Hi, my name's Viv. I'm the mother of Sophie, who is 18 months old. She had bilia atresia and followed up with a liver transplant when she was just short of seven months old. And I'm Andrew Day. I'm a paediatric gastroenterologist working here in Christchurch. Cool. Okay, so our first question is, what is bilia atresia? Yeah. So the liver sits up here on the right side, and it's connected to the body in a number of different ways, in terms of blood vessels going in, blood vessels going out. There's also a tube that comes from the liver down to the bowel, where the bile that's made in the liver gets transported.

In bilia atresia, that tube, the common bile duct, becomes completely scarred and blocked, so that the bile can't leave the liver and get to where it's meant to be in the gut. And what that leads to is progressive scarring within the liver that eventually stops the liver working, impacts upon how the liver's working. And in addition, bile's really important in terms of how we break down fats, how we get nutrients from our food. So when bile's not coming through to the gut, then that impacts upon absorbing fat, absorbing vitamins, and then upon baby's weight. What causes bilia atresia, and how common is it in New Zealand?

So it's not completely clear exactly what causes it. There are some congenital forms where there's changes that happen during pregnancy or when the baby's in the womb, but in most cases it begins soon after birth. And it's probably a combination of the immature immune response in that part of the body. Yeah. Perhaps triggered by viral infections, because there are some studies that show seasonal changes that go along with viral infections. And there's probably some genetic components that again we haven't fully understood. So the exact cause isn't known, but it's a combination of those different factors.

And probably somewhere around 12 infants in New Zealand are diagnosed per year. It varies a little bit from year to year. Yeah. So, bilia atresia is more common in some parts of the world than others. So in Taiwan and some Asian countries, for example, it's a bit more common. In New Zealand it's more common in Pacific people from the Pacific Islands, and people of Maori extraction, and a bit less common in Europeans. So what would the main cause of treatment be when Sophie was diagnosed with bilia atresia? Yeah. So, bilia atresia usually presents with jaundice, and it's a particular pattern of jaundice.

And then along with that, the stools are pale. And the stools are pale because the bile's not getting through into the gut. So when we're seeing a baby with jaundice, with pale stools who isn't growing very well, we have lots of concern in terms of biliary atresia, or perhaps one of the other things that can cause that pattern. Yeah. So there's a whole lot of tests that we do initially in that type of jaundice to work out the cause, and some of those causes of that type of jaundice are things that need immediate help. Some of them are things that get better by themselves, but we do a whole lot of tests, ultrasound, blood tests, urine tests, and a liver biopsy, all fairly promptly so that we get the answers as soon as possible.

And that's a really important part of things. And that's a message over the last years that we've been trying to promote more and more to midwives and GPs in terms of responding in the scenario of a baby with jaundice and pale stools to respond and refer early, because it's really important. Cool. Which is what happened to us, isn't it? And upon diagnosing biliary atresia and excluding the other causes. Yeah. Then the next step is an operation to bypass that tube that's scarred, so it's bringing a part of the bowel up to the liver, so the bowel can then drain through that segment and back into where it should be, and relieving the pressure or the impact upon the liver in terms of the scarring.

Cool. And that's called a Kasai procedure. It's a fancy name called portoenterostomy, but it was designed initially by a Japanese doctor. It was designed by a Japanese surgeon some decades ago called Dr. Kasai. And although it's been perfected and optimized a little bit over time, essentially it's the same procedure, but it's taking away the part that's scarred and then connecting up so that the bowel can drain. And upon diagnosing biliary atresia, that next step of getting the operation, again, Timeliness is really important. So we want to make the diagnosis and get on that next step as soon as we can.

2. Biliary Atresia - The operation, the recovery times after the Kasai and the importance of nutrition

And so when Sophie had her Kasai, she had to go to Auckland in New Zealand. It's the only hospital that does that operation, has surgeons who perform that operation. And so when, after she has the Kasai, what are the short and medium term kind of recovery times, especially having to stay up in Auckland if we're like ourselves and don't live there? Sure. So first of all, just in terms of, so in New Zealand, there's an understanding that this procedure is done in one location, i.e. Auckland. And that's because there's lots of data showing not only is the timeliness important, but also the experience and the expertise of the surgeon that's doing the procedure is really important to optimize the outcomes.

So someone that does a Kasai once every 20 years is going to have way less successful outcomes than someone's doing 20 a year, so to speak. And there's lots of, as I said, there's lots of data internationally to demonstrate that very clearly. The throughput and the experience is really important. And even if we were to look at international standards, New Zealand might still be considered as a sort of medium-sized centre, so to speak. But having it done with one group of surgeons in one location in New Zealand is really important. And in some parts of New Zealand, that initial workup in terms of could this be biliary atresia might happen in Auckland.

In the South Island, that would typically happen here in Christchurch. But then upon making the diagnosis, then yes, we would get up and get the operation organised. And then usually it would be a period of two, maybe three, sometimes a little bit longer, weeks up in Auckland. So not just a few days. And what we're looking for after the operation is to see, is the jaundice disappearing? Is the jaundice going in terms of the colour, but also the numbers in terms of the bilirubin when we measure in the blood tests? And ideally, we'd want complete drainage, i.e. that the bilirubin comes completely back down to normal, the stool becomes pigmented, and baby's doing well.

And in some cases, it doesn't drain at all, so nothing happens afterwards. And in some cases, it can be in between in terms of partial drainage. So the bilirubin might come down from 100 to 60, or come down to 40 or something, but not completely normalised. And that initial phase is really important in terms of assessing the response and the outcome to the operation. Perfect. That's exactly what we went through, isn't it, Sophie? I might just add one thing, Viv, as well. So in that initial phase, whilst we're waiting for the workup, for the drainage, we're waiting for the delivery. We're waiting for the delivery.

We're waiting for the test to come back, waiting for the Kasai. And even in the initial phase afterwards, nutrition is really important. So we would typically optimise the type of formula, the type of fat that the baby's getting to make it easier to absorb, so that baby can get the calories and the nutrients that it needs. And we'd also start on a whole lot of different vitamins, as well, again, to optimise the absorption of those vitamins. And that's vitamin A, D, E, and K. And that initial response in terms of nutrition and how the weight's going is really important in terms of the outcome, not just then, but also medium term as well.

Fantastic. Andrew, you've just answered my next question, which was, how does having biliary atresia affect feeding and nutrition, especially in the early months of a child's life? So that's great. And we can't say too much about the importance of nutrition. In this case. Yes.

3. Biliary Atresia – Signs or symptoms to watch for after the Kasai, and the outcomes

So after she or any child has had the Kasai operation, we're still kind of, we're always watching to make sure she was staying as healthy as possible, because I know that there are potential, increased potential for side, not side effects, but infections and kind of risks of having the Kasai. Yep. So can you just talk us through what those extra risks are, and following through with that, what are the signs and symptoms that as a parent I should be looking out for to know to bring her back into hospital after the Kasai? Sure. So in the weeks and months after a Kasai, the, it's really important in terms of that monitoring and the follow-up.

And that includes how baby's growing, how baby's feeding, how the vitamins are going and options. Optimising the levels, optimising the dose to correct the levels and ensure that things are doing okay. And obviously with the bilirubin, making sure it's coming down and stays down. With the Kasai, we've changed the anatomy, and we've taken away one of the defences, if you like, in terms of bacteria entering the liver. So essentially now the bacteria can move up that part of the bowel that's been used for the drainage and enter the liver. And there's a condition called ascending cholangitis. And we worry about that.

So for the first year after a Kasai or after any episode of infection, we'd use preventative antibiotics that can help in terms of stopping that problem. But in addition, if a baby presents following a Kasai with a fever or an increase in jaundice or an increase in some other liver tests, we would treat quite aggressively. In terms of ascending cholangitis. And what I mean by that is typically that might mean two weeks of IV intravenous antibiotics, along with an ultrasound and blood tests and monitoring and making sure things are responding. So in any baby that's had a Kasai that has a fever, if we can't explain it because of a sore throat or COVID or the flu or sore ears, we would want to treat for ascending cholangitis until we were proven otherwise.

Obviously, that has the short-term implications in terms of baby feeling not well, but one episode of cholangitis, a further episode increases scar tissue within the liver, and then can compromise how well bile leaves the liver into the gut and accelerate the need for transplant, for example. Cool. Thank you. Thank you. Understood. So yeah, you obviously talked about those are the signs and symptoms that were kind of non-negotiable to bring you back into the hospital. Are there any of those, well, are there any signs or symptoms that would be considered urgent that would need to call an ambulance?

I mean, probably not in terms of calling an ambulance. But if the baby that's had a Kasai has a fever, it's really important to be seen that day. Okay. As soon as possible. But not within minutes. Okay. It's not immediately life-threatening, but it is really important to respond. It's not something that one would wait at home until the weekend's finished sort of thing, but to head in. To help facilitate that, we do have care plans, what we call blue cards, which means that babies that are in this situation can front up directly to the Children's Emergency Centre, not have to go through. They don't have to go through other steps to get seen and assessed.

Excellent. And usually within that care plan, we would emphasise that we're worried about a temperature, and this would be the steps to consider and to do at that time. So we talked about, yeah, the liver scarring over time after the Kasai and things that can accelerate that need for transplantation. So I guess from a parent's point of view, being told we have this Kasai operation is a temporary fix, essentially. What are the chances that the Kasai will work? Sure. Is there odds? Is there any? What can we hold onto for hope that it's going to work or not? Sure. So when the Kasai works well, so maybe a third of situations, the bilirubin within a few weeks has come down to normal, and in that situation we'd be a little bit more relaxed, but we need to continue.

We need to continue the monitoring and the assessments that we've mentioned. And in some of those cases, some of those children, they would grow up there. I just saw a 13-year-old last week whose Kasai drained early, and he's still doing well. When I was training, there was a young lady in her early 20s who was having a baby, for example, and the Kasai was working fine. But it's really important to continue to monitor and make sure it's continuing to work well, because there does remain the prospect that there'll be issues, and that a transplant will be needed. In the worst-case scenario where there's no drainage and the bilirubin doesn't change at all, we're going to be even more anxious, and follow, monitor just that little bit more closely, because we'd be concerned that in that situation the baby will need to have a transplant early.

Okay. Maybe at eight months, maybe at 12 months. So it's looking out, so it's monitoring in terms of all the basic things, but in addition looking for things that suggest that the liver is compromised, and that arrangements for a transplant will be required. In the in-between group, which is probably again about a third of situations, we can be slightly relaxed in between in terms of monitoring. But looking out for worsening and for changes, and it might be that in that scenario, baby might need it, or the young child might need a transplant when they're three or four, not when they're eight months or 12 months.

4. Biliary Atresia – What can parents do and the importance of communication

So, we're wanting, as a parent of a child with biliary atresia, we're obviously wanting to hope that it can last as long as possible, delay needing a transplantation until she's bigger, stronger, and all those things. So, is there anything that we can do as parents of a child

with biliary atresia to improve her quality of life or essentially encourage her to keep her native liver as long as possible? So, I know we've talked about nutrition and presenting her to the hospital as soon as possible when there's any signs of ascending cholangitis. Is there anything else that we can do? So, ensuring that the little one is growing well, protecting in terms of the preventative antibiotics for cholangitis and treating appropriately, responding appropriately with fever or a change.

In some cases, we use medicines that could help in terms of helping to drain and keep things moving through the liver. There are other medicines that are coming and being assessed that may have a role in terms of helping to get drainage or to keep drainage early on, but they're not available routinely. I think the... I don't... I can't think of anything else particular from a parent's perspective, in addition to those things we've talked about. It's being aware and looking for changes and responding appropriately. Making sure she's growing and feeding her when she needs to. Yep. Cool. I think as a parent with a child with a chronic condition, you try to do everything that you possibly can that's in our control, because there's so many things that are out of our control.

So, I think that was a question we would ask ourselves often to see if there was anything we could do to, you know, optimise her future. So... And I guess, Viv, if I can just interrupt. Sure. I guess one of the things, and you may be able to add to this, is when things are unclear and when there's uncertainty, sometimes that can make people more worried. And it's really important to be able to express those concerns, whether it's to the paediatrician or to the nurse that's linking in or sometimes with a psychologist or social worker. There's other people that may be important in that team approach as well.

So, it's really important to be able to express and let people know of those concerns and worries. And whilst there may not be anything... ..that parents could do differently, it's really important not to sort of stew and worry, but to keep the team involved. I think as a parent in this situation, it was nice always being informed of what is happening and what could possibly happen, just so that we were kind of always aware of how the journey could go, because it could go in so many different ways. And I think... ..having a really great medical team, which we did, always keeping us up to date of what those things are and not kind of keeping things hidden.

I think there were times that we might have felt that we weren't being told the full picture, but in hindsight, that was probably how we felt at the time. In hindsight, it was because it was... ..everything was a little, not uncertain, but, you know, we kind of just had to see how things played out. And... ..yeah, I think just uncertainty over anything makes anyone quite anxious, but keeping up the information and the education was really helpful.

5. Biliary Atresia - What are the pros and cons of having a liver transplant_

So for Sophie, her Casai didn't, essentially wasn't as successful as we wanted it to be as early as possible. So then we faced the kind of question of what are the pros and cons of having a liver transplant early in life? So she had hers just under seven months old. So having it as a young baby, are there any pros and cons compared to another biliary atresia child who might be having it a bit later? Sure. So part of it reflects how much the liver is compromised, how well it's working or not working. So during that sort of phase, it's looking out for growth, it's looking out for what's happening with the bilirubin, it's looking out in terms of the liver.

So the liver does more than 200 different jobs, but some of those are much more important, particularly in terms of how the liver's making proteins. And there's a whole series of things that can happen when the liver fails, and that can happen in an adult or a child. But we want to try and get the right time to intervene in terms of moving ahead with the transplant before the baby or the child becomes too sick. We don't want to make the decision too early, but we also don't want it really important. And secondly, we don't want to leave it too late in terms of when the baby is very sick and the liver is severely compromised and under-performing.

And it's always trying to get that right balance. And that's why following, monitoring very closely, checking in with growth and looking out for other things in terms of how the liver's working or not working is really important. Okay. In general terms, when we are concerned that it's a bilirubin, it's a bilirubin, it's a bilirubin, it's a bilirubin, it's a bilirubin. Yeah. Yeah. It's inevitable that there's going to need to be a liver transplant, then we would arrange for a transplant assessment. And much like the CASAI procedure being done in one centre, liver transplant for children and adults for that matter is done in one centre, i.e.

in Auckland, and it's the same group of surgeons. And so a transplant assessment is essentially a week in Auckland where there's a whole series of tests that are done. There's a whole series of discussions and meeting different people, explaining this, that, and the other. And so part of that is to make sure that a transplant, moving to head to the transplant is the right thing at this point in time, and also making sure that there's nothing that's going to compromise the transplant in terms of a heart problem, or a lung problem, or pneumonia, or some other issue. Or having cancer. Yeah. Yeah.

Yeah. Well, having COVID, yes. And at the end of that week of a transplant assessment, then there'd be a team discussion, and yes, the decision would be made to list the child for a transplant. So it's not that at the end of that week, they automatically get their new liver. And then it's going on to the transplant list in terms of being ready to... to proceed for them.

6. Biliary Atresia – What it was like as a parent and the organ donor process

So, following on from the transplant assessment side of things that I just mentioned, could I ask your perspectives of that process in terms of there must be lots of emotions and lots of concerns during that time? Yeah, I think for us it was really daunting when we knew that she was up for a liver transplant, but equally almost a bit of a relief that we were there because we'd gone through the past few months of kind of watching her bilirubin going up, knowing she was getting sicker. We knew that it was going to be the inevitable, and so when we got to the point of the assessment, we knew at least things were in motion.

So that was actually kind of probably a bit of a relief. It was a very intense week of learning about what will happen. It's been during the transplant and what life looks like after the transplant, but equally Sophie's dad and myself, we quite, like I kind of said earlier, we like being informed. We like knowing things, and I think it made us feel a lot better that there are lots of children actually out there. There's been children before Sophie who have been through this process, and they have been okay. They're living normal lives now, and there are measures in place to help avoid the risks and all that kind of thing.

So yes, it was daunting, but interesting. I don't know if interesting is the right word, but yeah, we knew this was what our life was going to look like now. Again, it was out of our control, so we might as well embrace and make the most of it and make the best of it. Mm-hmm. So learning that being a live donor as an option was really helpful because I think in our heads, you hear about a transplant, being on a transplant list, you need to wait for an appropriate organ to be available. And I know with certain organs, you do sit on a list and have to wait, but with the liver, we learned that we don't actually have to wait.

Yeah. We don't actually have to wait for a deceased liver that's the right size, that's the right plumbing, that fits Sophie, that we can go through a live donor option. And that means that we wouldn't have to do the waiting process as long. So going off of that, can you explain the organ donor process in New Zealand to us? Sure. Of course. So after that transplant assessment and then the baby or the child is then listed, so on the transplant list, there might be two. There might be eight children on the list. It varies a lot from time to time. And they're sort of stratified in terms of their clinical priority.

So the sickest children will be at the top of the list, and those that can wait a bit longer in terms of how well they are will be towards the lower part of the list. And then, yes, there's two ways to get a liver. One is from a deceased donor, and it's not that the deceased donor needs to be the same sex or the same size. But generally, in most cases, the liver needs to be matched by the blood groups. So there needs to be some compatibility. And in some cases, the donor liver will be divided into two. So the left lobe will go to one recipient, and the right lobe to the other recipient. And sometimes it's trimmed down to size or reduced.

Okay. To fit in to a child. So having one lobe, for example, in a smaller child might be just the right size for their abdomen to fit in. Okay. And then what happens to that liver as Sophie grows bigger? Does, you know, what happens if it's a little bit too big or, yeah, I guess the question is what happens down the line when it's someone else's liver in her? Sure. So. Well, maybe I'll answer the other question first. The other way to get a transplant is from a living donor. And in that case, the donor, it may be a family member, it may be a friend, it may be some other relative, grandparent, for example, in that case the donor needs to go through an assessment to make sure that their anatomy is okay, that there's no surprises for the surgeons, to make sure that they're not being coerced, that they're making the decision for the right reasons.

So that's an assessment in Auckland as well, in terms of a donor assessment. And then the left lobe of the donor is taken and transplanted into the recipient. The liver is very good at regenerating. Okay. Yeah. So even in someone that's had the left lobe donated to someone else, their liver will increase in size over time, whilst in some cases in an infant or a child, the liver that they receive might be big initially, and very uncommonly there will be difficulty closing the abdomen for that reason. Okay. So, obviously that improves over a little bit of time, but subsequently the liver continues to grow with the child.

So unlike many other organs, or any other organs, the liver is very good at regenerating. And so again, in a child that's seven months or eight months that receives the liver, it's not like the liver if it comes from a 30-year-old, for example. Okay. Yeah. It will then take on the form, if you like, of the age of the recipient. Wow. That's very cool. Amazing. And then, so once the liver transplant does happen, what does the short and medium-term recovery look like? That, again, kind of similar to the Kasai question, for those of us who have to travel to Auckland, how long will we be away from home, how long will she have to be monitored closely in that centre?

Before being able to come home? Sure. So, the concerns following a transplant include all the plumbing, so it's connecting the bile, it's connecting the blood vessels, and some of the initial concerns are in terms of how those connections are. The next sort of line of concern is infection, so it's a big operation, there's risk of infection locally or in the wound or whatever. And then there's the further concern is rejection in terms of, so the liver, as mentioned, needs to be matched in terms of blood groups, but there's still a risk of the body rejecting that liver and having an immune response that then attacks the liver.

So initially there's a number of medications that we use to protect against infection, there's a number of medications that we use to protect against risk. Okay. There's a number of medications that we use to protect against rejection, and it's following the liver tests, it's

following how things are working very closely initially. So initially the baby might be in intensive care for two days, three days, or it varies a bit, and then on the ward for a period of time. And then usually for at least six weeks the baby would be in Auckland because they'll be being seen daily, every second day, progressively less frequently, checking liver tests, checking how the liver's going, checking how everything else is going.

And then come back to their home centre. And how long that process depends a little bit about how well things have been, but often babies coming back to Christchurch like yourselves might be six or eight weeks, but in some cases that might be 12 weeks in Auckland. So it does depend a little bit upon how things have gone for that child receiving the liver, but also where they live in New Zealand as well.

7. Biliary Atresia - What we learnt as parents, advocating for one's self

Through the last 18 months of us going through all of this, I think a big thing that we've learned as a family with a child with a chronic condition is just learning to advocate for ourselves, especially as parents who aren't medical professionals. There are times where we're not sure quite what you might have explained to us or there have been times that we've presented at the emergency department and we maybe don't necessarily agree or understand with the actions that have been taken. Is there anything you can share with us about what we can do to advocate for ourselves if we're not sure or if we don't agree?

Okay, so I think the really key things are talking to the team that's involved as the key team. In New Zealand there's also the option to liaise with one of the liver nurses in Auckland as well as another point of contact, but linking in with the local team is one of the really key things. The hospital does have advocacy services and for some people having that external person to help make their voice heard, it can be helpful. But I guess the really key thing is to be able to express and to voice concerns and voice what people are worried about. Okay, yeah, I think one of the things I discussed earlier with Sophie's dad was, especially in the early days when you're explaining, you know, what bilaterality is, it's a brand new thing for us and not really being maybe necessarily able to process what that all means for us the first time you tell us about it.

Is there any ways that we can get follow-ups or explanations maybe to be repeated or written out if that makes sense? Or is that just something that we just need to be confident to ask the team for? I mean, I think people will differ in terms of their confidence to ask, but I think the really important thing from our perspective is to be aware that this is a whole new field, a whole new lot of things that people won't be aware of. And as you know, I often draw pictures and try and explain things in that fashion and leave the pictures and then we can come back to them and add or clarify.

And I try to make sure that when we're talking to people, we don't just look at a picture, and then we're thinking about, talking the second or the third time but there's lots of time for questions or clarifications and that's a really important part of things in terms of this is a learning curve and it's not going to be an instant understanding and full comprehension of what might be happening now but what the implications of that will be as well okay anything another just a key message or key question for families going through this is just even at more of a base level when we come in for things like blood tests or you know there's something that we're waiting to hear from so for example in those early days when we were running lots of tests and not waiting for results what's a good way to contact you well contact our team I think there was something that we weren't really sure of do we call do we email is there what's the means of communication sure I mean that will differ a little bit from center to center and from person to person but in essence there are lots of ways to get in touch into contact so sometimes it can be through the nurses for example in the outpatient setting we have historically had a nurse that's had more of a role in terms of linking in with families like yourselves in the community so one of the outreach nurses having a responsibility and a team role and sometimes that can be an important way to contact so through the outreach nurse or the nurse that's working in the community with with people with families and then through the junior staff and the team can be another way to contact as well and then yes so sometimes email is a good way to email through the paediatrician or paediatric gastroenterologist is another great way to contact as well I think yeah I think as a family we're never sure what the follow-up is you know when we went through a phase where we weren't sure when something happened do we wait to be heard or do we make contact so I think again it was just communication with the team of what works best with how we work