

New Zealand Liver Transplant Unit

Paediatric Liver Transplantation

Parent Guide



Your Shared Care (local) doctor is:

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Your Shared Care (local) nurse is:

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Your Starship doctor is:

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Your Starship nurses are:

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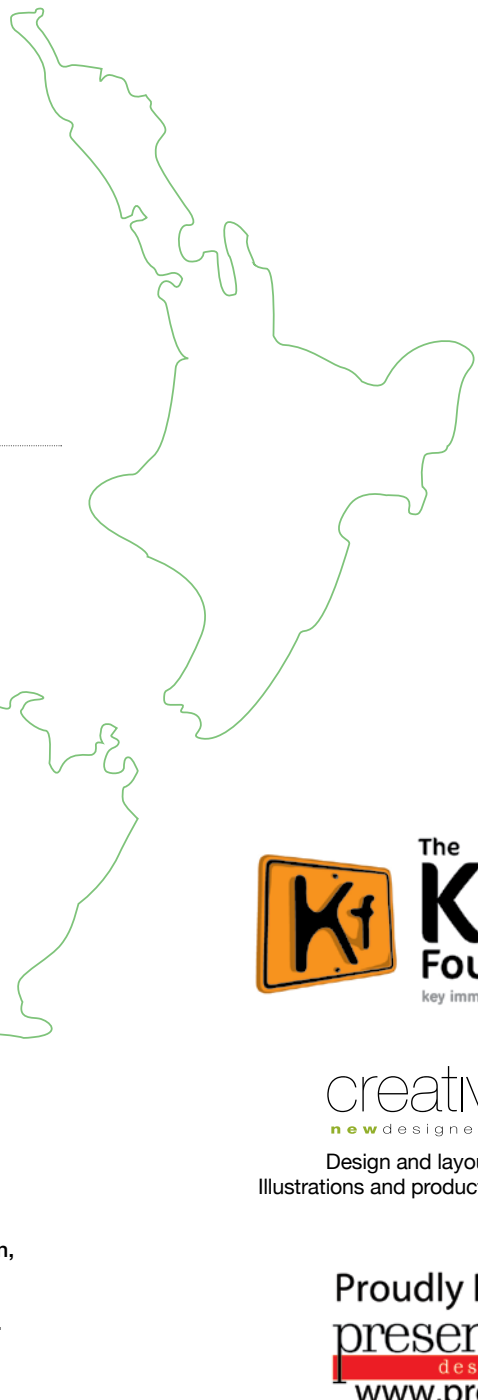
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Information provided in this parent guide is a guide, it is not intended to replace qualified medical or professional advice. For diagnosis, treatment and medication, you should consult your child's medical specialist team.

Every effort has been made to provide the most current and relevant information. Updates and changes to practices will be made from time to time.

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Design and layout by Erin Holmes
Illustrations and production by Tracey Liu

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Acknowledgments

.....

Stephen Munn
Peter Johnston
Liver Transplant Surgeons

Margaret Johnston
*NZLTU Senior Liver Transplant
Coordinator/Nurse Practitioner*

Karyn Sanson
Clinical Nurse Specialist CNS

Contributions from:

Adam Bartlett
Transplant Surgeon

Simon Chin
Helen Evans
Stephen Mouat
Jonathon Bishop
*Starship Gastroenterology
Consultant Team*

Caroline DeLuca
Paediatric Pharmacist

Kaiatawhai Team
Kim Herbison
Clinical Dietitian

Contributions & Edit completed by:

Samantha Sutherland –
*Mother of Transplant recipient Lara
Sutherland*

Cate Fraser-Irwin
Clinical Nurse Specialist CNS

Professor Ross Shepherd
Hepatologist

Jonathon Bishop
Hepatologist

Thank you to our families who most generously contributed photos and their words to support families who are now just beginning their own transplant journey. We are immensely grateful for your kindness, honesty and hope. Also a thank you to the 26B nursing and medical staff who put their all into managing the care of our transplant patients pre and post transplant.

Cate Fraser-Irwin

Simon Chin with Baby Alice



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Embarking on the journey of liver transplantation is the start of a lifelong relationship between the liver transplant service and the child/young person and their family. We hope to provide you with the information and tools to negotiate the complex health care system as successfully as possible and build a positive relationship with you based on trust.

If things seem to be getting on top of any family member, ask for help. There is always someone who can talk to you to try and help you get through this journey in one piece!

“It is not easy....it is hard for all the family....but you get through it.”

– Donatella

(age 15yrs transplanted for acute liver failure 2011)



Some of the team members you will meet: Karyn Sanson, surgeons Peter Johnston and Phil Morreau, Helen Evans, Cate Fraser-Irwin with patient Maria

“A really rough ride...such a roller coaster but when you look at him now...10yrs post transplant...it was so worth it!”

– Angela, mother of Naziah

Introduction

This guide aims to walk you through the process of your child being assessed for a liver transplant, the waiting period before a liver transplant, having the liver transplant and then living with their new liver. The guide is written in sections and we will spend time with you as a team going through its content and answering your questions along the way.

Every question you have is important to us, so please never be afraid to ask. Ask as many times as it takes for you to feel you understand what is happening to your child. As much as possible, it is important to convey this understanding and confidence in going forward to your child and include him or her in the process to the level of his/her understanding.

Children with liver failure are one of the most medically fragile and complex groups of children cared for at Starship Children's Hospital. They require close monitoring and carefully timed interventions to ensure the

best outcomes possible. Deciding to undertake a transplant and then waiting for a transplant in this situation are perhaps the hardest stages of the whole process. In addition, liver transplant surgery in children is recognised as one of the most difficult of all surgical operations, and the after care is at times complicated.

To achieve the best outcomes possible, good teamwork is paramount. You are part of the team! We encourage you to contribute your observations and say your piece. Together, we aim to keep you/your child in as good a state as possible while waiting; to match and find a good donor; to carry out the transplant to the best of our ability; and to monitor for and manage any problems which might arise after the transplant in a timely manner – with the ultimate aim of achieving as normal a life as possible for your child and your whole family.



Oliver with his sisters

“If I could sum up our story in a few sentences it would be terrifying, life-changing, supportive and amazing. Ollie went from acute liver failure to doing roly-polies in 10 days. Ollie's sisters, himself and I were supported by so many different people at Starship and IDFNZ from the start and it keeps on coming! Amazing. And he's soooo proud of his scar.

My super hero.”

– Chloe, mother of Oliver (5)

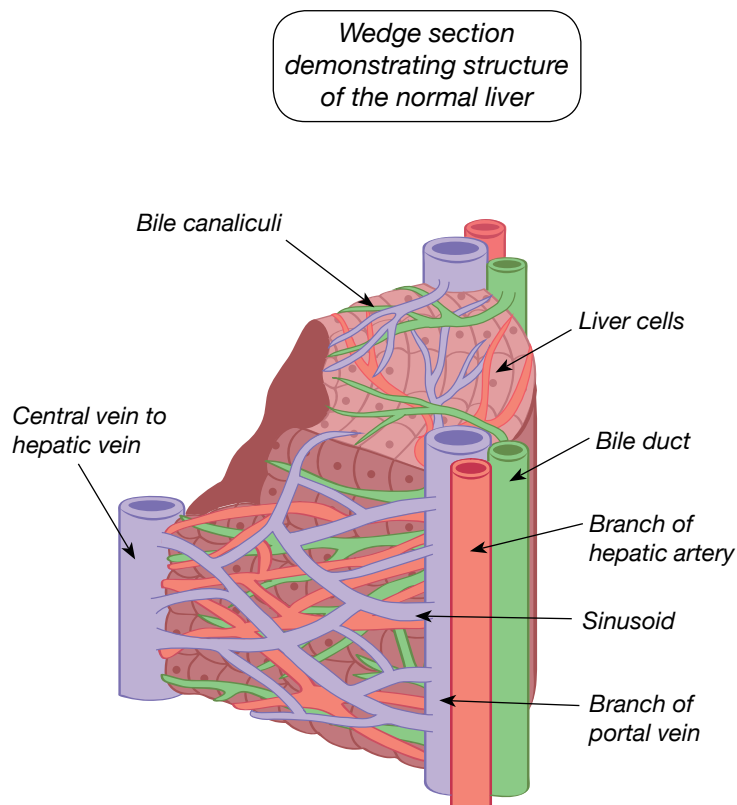
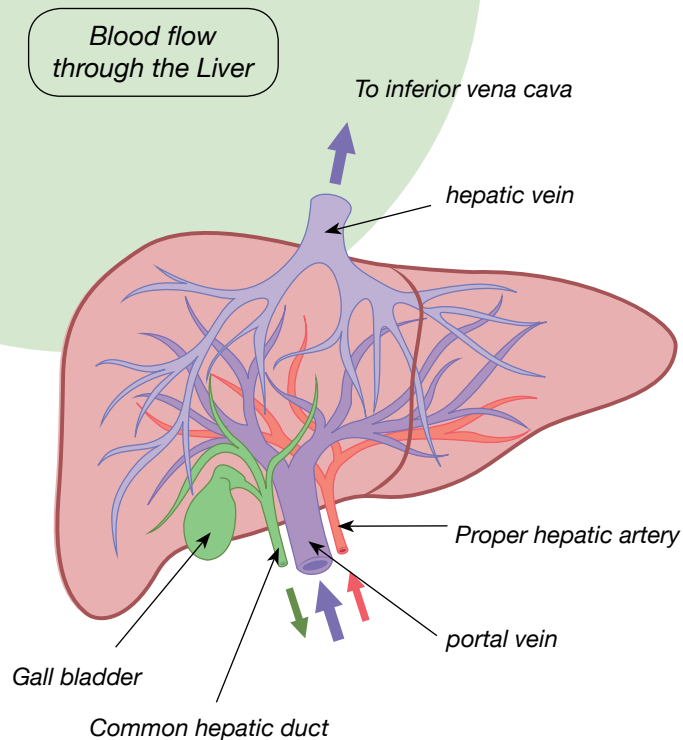
What is the liver?

The liver is a vital organ (you can't live without it!) and is the largest glandular organ of the body. In health, it is reddish brown in colour and lies on the right side of the abdominal cavity beneath the diaphragm. It is divided into two lobes of unequal size and shape, divided by a ligament (falciform ligament) and, fortunately, by virtue of its plumbing (the blood vessels going to and from the liver, and the bile ducts taking bile to the intestine), it can be surgically divided into segments (eg for partial liver transplant—see later), and has a remarkable and unique capacity to regenerate itself to a normal size if some segments are removed.

The plumbing is important!

Blood is carried to the liver via two large vessels called the **hepatic artery** and the **portal vein**. The hepatic artery carries oxygen-rich blood from the heart and lungs via the aorta. The portal vein is the source of 75% of the liver's blood supply. It carries blood containing digested nutrients from food from the small intestine and breakdown products of blood from the spleen. Both of these large vessels subdivide in the liver repeatedly, terminating in very small blood vessels, called capillaries. Each capillary leads to a lobule. Liver tissue is composed of thousands of

lobules, and each lobule is made up of liver cells, the basic metabolic cells of the liver. These liver cells perform all the functions of the liver and are lined up so that their secretions can leave the liver, either into the bloodstream via the hepatic vein, or in the bile. The **hepatic vein** transports deoxygenated blood from the liver to the heart and remaining glucose and amino acids to the rest of the body. The bile is secreted by the liver and is stored in the gall bladder.



What does the liver do?

It's complicated! ...Some people think of the liver as the body's power station (supplying energy to the body), but it is also a chemical plant and a waste station.

Basically, your liver processes blood. This involves, breaking down stuff (the nutrients and chemicals your blood carries), storing stuff your body might need, making new stuff your body needs, or getting rid of stuff the body no longer needs. To do these things, as you've seen, the liver has two blood supplies (one bringing oxygen and one bringing nutrients), and two ways to get stuff out (either via veins to the heart, or via bile to the intestine). But why exactly does your liver need the oxygen and nutrients supplied by your blood?

Well... there are over 500 vital functions, though these come under three headings:

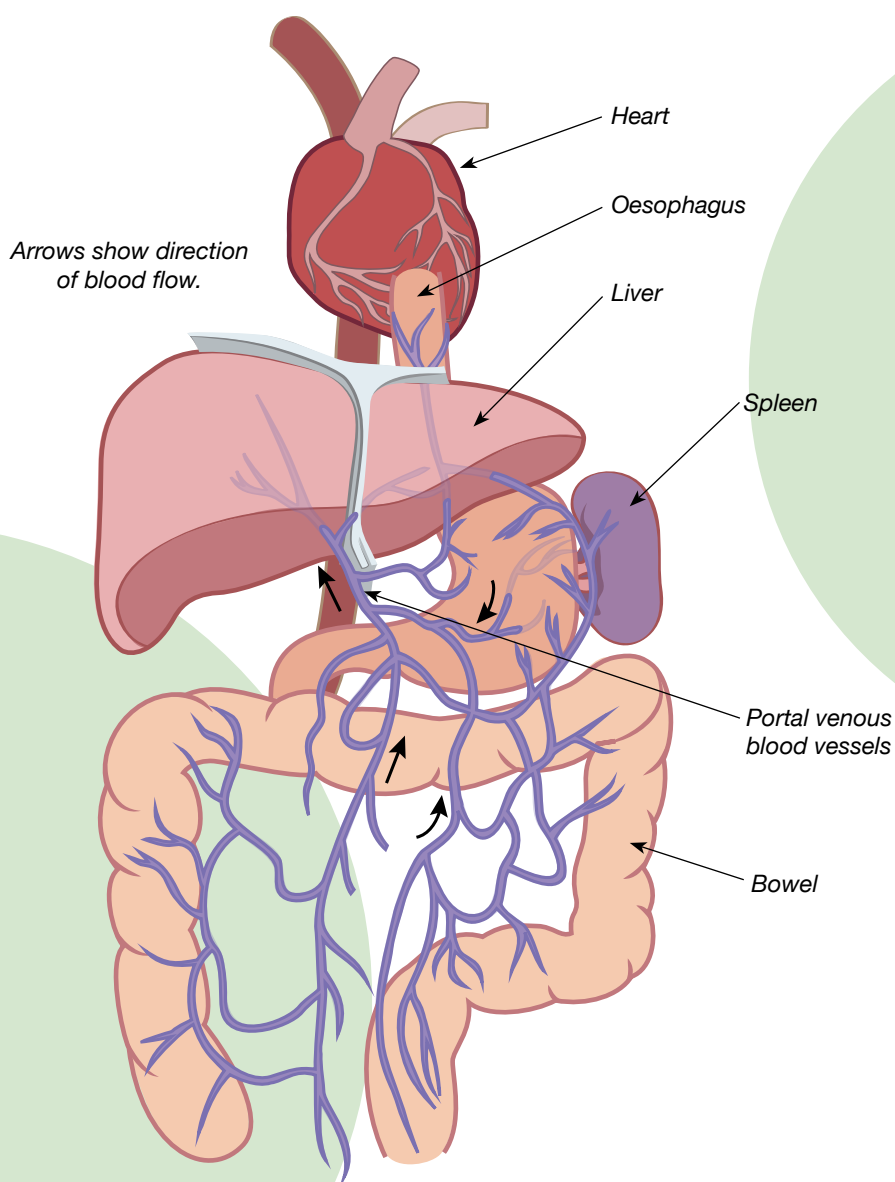
1. Production and transportation of bile. Bile is a yellow/green thick liquid made by your liver, which is transported to the intestine via the 'biliary system' (see Figure),. One role of bile is to remove waste products (such as bilirubin and toxins) from the liver. Bile also contains useful products that help break down and promote absorption of fat from our food. Some bile substances (eg bile salts) are re absorbed back to the liver for re-use. So even though bile is partly made up of waste products, it certainly doesn't go to waste!

2. Storage and conversion of nutrients necessary for energy.

Energy is stored in the liver as carbohydrate, fatty acids, and certain proteins.

3. Production of proteins vital for normal bodily function, particularly for fluid balance, blood clotting, immunity, hormonal balance, growth and nutrition.

Normal portal venous blood flow



How do we test for liver problems?

Scans & X-rays (these test the liver anatomy and plumbing)

- Liver ultrasound (which can detect abnormalities of the size/shape of the liver and can see whether the blood vessels and bile ducts are ok).
- Cholangiogram (direct x-rays of the bile ducts)
- Angiogram (direct x-rays of the blood vessels)

Blood Tests

'Liver function tests' or 'liver numbers' are blood tests used as a guide as to what is going on in the liver. Each result has a number, and the laboratory provides a 'normal range' to the test, which can vary with age and sex, showing whether or not the number is within the normal range. These are:

- Alanine aminotransferase (ALT)
- Aspartate aminotransferase (AST)
- Alkaline phosphatase (ALP)
- Gamma glutamyl transferase (GGT)
- Bilirubin
- Albumin
- Clotting studies: prothrombin time (PT) or international normalised ratio (INR)

High ALT and AST levels usually indicate some sort of inflammation of the liver cells. These enzymes are present in the liver cells, which leak into the blood stream when the liver cells are damaged for any reason.

Alkaline phosphatase (ALP) and Gamma glutamyl transferase (GGT) are enzymes found mainly in the bile ducts of the liver, but ALP is also found in bone. Increases in ALP and the GGT may indicate something is going on in the biliary system, for example, where bile is not properly getting out of the liver because of obstruction (blockage) of the bile duct. GGT is tested with

ALP to make sure that ALP increases are coming from your liver.

Bilirubin (formed from haemoglobin and the main pigment in bile). When bile cannot be produced or flow from the liver for some reason, the bilirubin spills over into the blood which causes jaundice (yellow eyes and skin).

Albumin is a very important protein produced by the liver that carries stuff around the body and helps maintain fluid balance. Low Albumin occurs in chronic liver disease, particularly if the disease is getting worse, but may be low for other reasons such as a lack of protein, for example in malnutrition.

Abnormal Clotting studies can occur for two main reasons in children with liver disease. Liver disease can affect the body's ability to absorb some vitamins from food. These include vitamin K, which is required by the liver to make some compounds that help the blood to clot (clotting factors). This problem can be solved by giving extra vitamin K as a medication. However, in very severe liver disease, the liver may be too sick to manufacture these clotting factors, even with extra vitamin K. If this occurs, the blood becomes too 'thin' and takes longer to clot, which may lead to easy bruising, or bleeding which takes a long time to stop eg nosebleeds, or after blood tests.

Common blood tests and their normal values

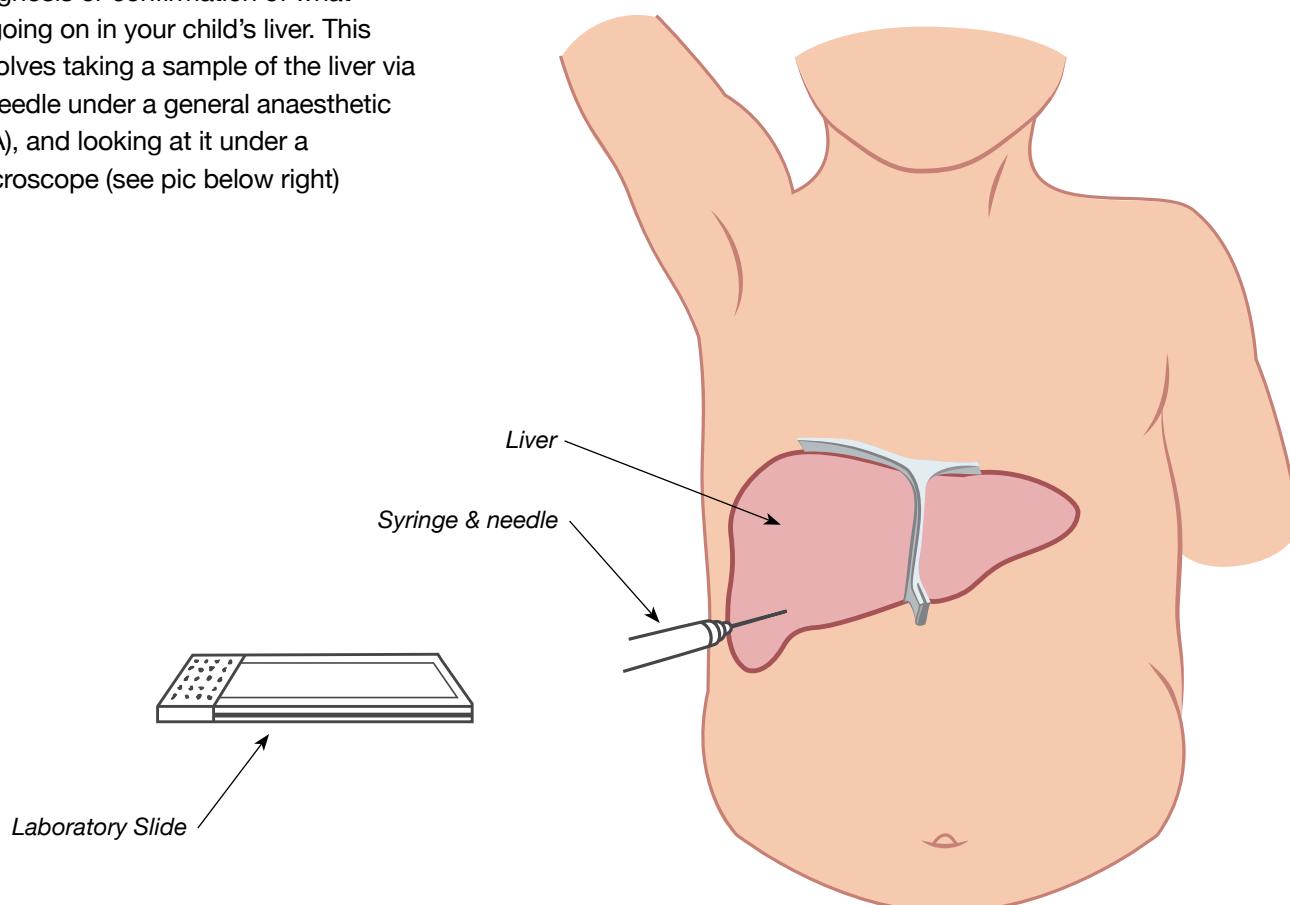
The table below lists the common blood tests used to assess liver function and their normal values.

Test	Normal Values—Babies	Normal Values—Older Children
Bilirubin—Total	0–24	0–24
ALP	80–350	45–250
GGT	0–50	0–50
ALT	<45	<45
AST	0–80	0–80
Measure of Synthetic Function		
Albumin	38–52	36–50
INR	0.8–1.2	0.8–1.2

(Adapted from Starship Éclair (clinical reporting system))

Liver Biopsy

This can give a more accurate diagnosis or confirmation of what is going on in your child's liver. This involves taking a sample of the liver via a needle under a general anaesthetic (GA), and looking at it under a microscope (see pic below right)



What are the reasons for liver transplant in children?

The commonest reason for liver transplantation in children is chronic liver failure, which may be caused by a number of conditions. When the liver starts to slowly fail your child may not grow as well as normal, or may develop complications such as bleeding from the gastrointestinal tract (variceal bleeding), build up of fluid in the abdomen/tummy (ascites) or repeated infections in the liver (cholangitis). Your child may become more tired or sleepy. These complications may occur despite improving nutrition or using medicines such as antibiotics or procedures to try to help. The liver is an extremely complex organ and, if it is very scarred, it may become impossible for it to perform all its essential functions.

The most common cause for chronic liver failure in children is biliary atresia. There are many other less common causes, including genetic, metabolic and auto-immune diseases.

Occasionally, a liver transplant is required because the liver fails suddenly (acute liver failure). This is often due to an infection, but sometimes drugs and metabolic diseases can also cause liver failure.

Rarely liver tumours are a reason for liver transplant.

Finally, liver transplant may also be required for some rare metabolic conditions. These are inherited

conditions, in which the body lacks the ability to breakdown or use various products of digestion. These conditions may not cause chronic or acute liver failure, but may have effects on the child's health which mean Liver transplant is the child's best treatment option.

History of Liver Transplant

When was the first liver transplant performed?

The first experimental adult liver transplant was performed in the US by Dr Thomas Starzl in 1963. He also performed the first liver transplant in a child in 1967. However, it was not until the 1980s that developments in surgery, immunosuppression (drugs which prevent rejection of the new liver) and intensive care had improved outcomes to the point where the procedure was no longer considered experimental. However, for children, there were not enough donors until the first reduced size (using segments of the liver) 'cutdowns' and 'splits' of adult donor organs were developed in the 1980s by Drs Bismuth and Houssin (1984) & Dr Pichlmayr (1998) in Europe, and



Dr Thomas Starzl

by Dr Russell Strong in Brisbane, Australia (1985), who also performed the first successful reduced size live donor transplant from a mother to her child (1989).

It turns out that part of an adult liver (eg segments II and III) can fit nicely into an infant or young child, and grow and work successfully indefinitely. Over time, increasing use of partial liver transplants, improved ways to preserve donor organs, and better drugs to prevent rejection have seen a marked increase in liver transplants in children. Now, more than 2,500 transplants are performed worldwide in children each year, of which about 70% are reduced size and 40–50% are from living donors.

In Australasia, the first liver transplants in children were performed in Brisbane in 1984, where reduced size and living donor transplants were early developments. During the late 1980s and 90's, New Zealand children requiring transplants were referred to Brisbane, which necessitated long periods away from home, returning to New Zealand once stable to be followed up by the paediatricians in their home towns. The New Zealand Liver Transplant Unit commenced adult transplants in 1998 and started transplanting children under 7 years of age in 2002.

How successful are liver transplants in children?

The success rate these days is very good and the majority of children lead fairly normal productive lives after liver transplant. The oldest person we know who had a transplant as a child (aged 4 years) is now in her 30's, has been to University, got married and had two children!

The international experience to date (inclusive of Pittsburgh in the States, and Kings College and Birmingham in Britain) reports more than 95% of patients survive more than one year, and around 87% by five years.

The New Zealand Liver Transplant Unit commenced service in 1998, with close to 500 liver transplants having been done to date for both adult and paediatric recipients.

Since the start of the Paediatric Transplant programme in 2002 and May 2012 we have performed 80 transplants in 77 children with close to 98% survival rate (1 year 98% survival rate, 5 year 96% survival rate). We are very mindful as a team that we have a small program and that this level of success is a fragile state.

Most children listed for transplant actually receive one, because of the various donor options available. However, serious complications of liver disease can occur while waiting and there is a small risk of dying on the waiting list. After transplant, 95–100% of patients are alive after 1 year, and around 90% after 10 years, so there is a small risk of dying of untreatable

complications after a transplant. Some patients (about 1–3%) do require a second transplant because the donor liver fails.

The vast majority of children who have had a transplant are well, often leading normal lives within 6 months of their transplant. However, they do require lifelong medication to prevent rejection and ongoing follow-up by the transplant team. Serious complications can occur, such as graft failure and infection, and these are usually, but not always, manageable. Other complications including rejection and bile duct plumbing problems are almost always treatable.

Naziah (Left) Liam (Middle) Lara and Blake (Right)





“ Trust in the medical team, they have the same goal as you – a healthy child. It’s scary but it does get easier – take it one day at a time. ”
– Kate Manson, mother of Alice.

“ I found it helpful to set up a caring bridge website – I could journal how our day went to keep family and friends updated without having to keep talking about how hard it was. People could post supportive messages which gave me a boost.

Also take mental health breaks – even if you can’t bear to leave your child – make use of offers of help or hospital grandparents and take a walk to have some time out – you deserve it and need it. ”

– Samantha
mother of Lara



“ At the beginning of Jordan’s journey it was like a natural disaster hitting us over and over again. We never knew when the next wave was going to hit us, the storm came and went on its own accord.

Everything was traumatic and unexpected. Eventually we learnt to ride the wave and dance in the storm. Transplant time was definitely the most intense time for us but with support from family, friends and others sharing a similar experience we made it through. Also seeing children and other families who were worse off than us on a daily basis helped



Jordan



me be thankful that Jordan was still alive and smiling. I always wondered when it was all going to end.

Today Jordan is the best he has ever been in his 5 years of life. He still has his ups and downs but at least now he spends more time out of hospital than in. My advice would be to ride that wave, expect the unexpected and learn to dance in the rain. Make the most out of a not so good situation and find a reason to smile every day. Be strong for your babies and they will be strong for you. Kia ora! ”

– Lakita,
mother of Jordan

Where does the “new” liver come from?

There are two possible ways for a liver (or piece of one) to be donated: Deceased Donation or Living Donation. To ensure the best outcome for your child's transplant, all potential donor options are assessed. The major considerations to getting the best possible donor include timeliness, size matching, blood group, availability, suitability, and of course consent from either the donor family (if deceased) or from the potential donor (if living).

Deceased Donation

The deceased donor liver usually comes from someone who has recently sustained an irreversible, fatal brain injury, such as from a road crash or a brain haemorrhage. His or her family have been given the option to donate their organs and consented to donate their loved one's liver, or other organs, to give life to someone else.

The donor will have been admitted to an Intensive Care Unit and placed on a ventilator (breathing machine). A number of tests are carried out to show that there has been a complete loss of brain function resulting in “brain death”.

The New Zealand and Australian Liver transplant programs have a cooperative sharing program of deceased donor organs. This is based on certain priorities such as the severity of liver disease in those waiting for a transplant, and matching of organs for size and blood group. An adult deceased donor liver can be used whole, or ‘split’ so that one larger section could be transplanted into an adult and the smaller segment given to a child, benefitting 2 recipients. A child

deceased donor liver, which is not as common, is given to a recipient child of a similar size.

Donor details are confidential. Although there is a natural curiosity about this, please understand that at no time will the transplant team reveal to you any details relating to the identity of your child's donor. You will be given the opportunity to write to the donor family if you wish when a reasonable period of time has passed. You will be given a copy of the guidelines developed by the donor coordinator service about this. (See the section later on Writing to donor families).

Live Liver Donor

“42% of children who have been transplanted since the beginning of the NZ paediatric program have received their organ through live donation.”

A live donor is usually an adult relative or friend aged between 18 and 60 years who gives part of his or her liver to the patient. The donor will be matched for both blood group and plumbing (size and availability of blood vessels) to the recipient. Both the part donated and the part which remains in the donor grow to normal size within 6–8 weeks of the operation.

The main advantages of live donation are that the transplant operation can be planned in advance at a suitable time for the donor, the recipient and the transplant team. Therefore, the waiting time for a transplant will also be significantly reduced.

However, it should be understood that the donor operation is a major surgical procedure and there are

some risks to the donor. Only certain patients and donors are suitable for a live donor liver transplant. The donor must be in excellent health and pass a rigorous evaluation. Live donor surgery is the only surgery done when a patient is well.

Potential live donors are evaluated and treated by their own multi-disciplinary team including a physician, surgeon and psychiatrist and social worker, who are separate to the recipient's transplant team. This enables the evaluation to be unbiased and allows anonymity to the potential donor should they wish to withdraw from the process.

If you, a family member or friend are interested in pursuing live donation as an option, the Nurse Specialist will provide you with a copy of the living donor guide. Once you have read this you are welcome to contact the liver transplant coordinator on-call via the to hospital switch board. They will discuss the process with you and make an appointment to complete the preliminary investigations.

The evaluation is not undertaken by the paediatric team. We will not be able to discuss with you any progress, or in fact know whether we have a suitable donor for your child until it has been finalised and confirmed with the donor. Your child will need to be formally listed before potential donors can contact the on-call liver transplant coordinator.

Parents' perspectives—liver donation

A Father's Perspective

After almost losing my son, (he was 3½ years old at the time) to acute liver failure, I am one who understands the importance of organ donation. These donors are not only important to the people who are in desperate need of a donor organ but also to the families of the sick and dying. I do not know what life would be like for me without my son, but what I do know is that losing him is something I would not cope with well.

I believe a son should bury his father not the other way around. Sitting in Starship hospital, PICU day after day waiting for a donor for a son who is in a coma would be the hardest thing I have done in my life. After sitting with him all day and night, getting a few hours restless sleep, waking the next morning



Sean, father of Liam who is now 10 years old and thriving

without the phone call to say a donor had been found was like ground hog day.

My family is one that had a happy ending. A school friend of mine was able to donate part of his liver to save my son's life. This was only after my wife and I were ruled out as donors because of being the wrong blood type. Had we not had close friends and family our son would have died. A little boy less than 2 years old was not so lucky. He died not more than a couple of months after my son's transplant.

To see my son today you would not know the hardship he has faced. He has started school, he's learning to swim and generally enjoying life like any other 5 year old. None of this would have been possible without an organ donor.

Being a Live Donor

(Mum, Jodie, for Isabelle aged one year and five days at transplant)

The decision to be tested as a live donor for my daughter, Isabelle, was an easy one for me and I was really, really pleased when I got the okay – it meant that we could go ahead with the transplant at a time that was best for her. Being the parent of a sick baby I felt completely hopeless and to be able to donate made me feel like I was actually helping bring her back to good health.

As a Mum I was worried about when I would be able to see Isabelle again after the surgery, and it was also really important for me to know that her Dad (my husband) would be there for her while I was recovering. My Mum came up from the South Island to be my main support person.



Isabelle's first swimming lesson



On the day of the transplant I was nervous, mostly about the outcome for Isabelle, but also excited that the time had come to get rid of her failing liver. I remember waking in CCU and being thrilled and relieved when I was told that it had gone really well. Both the medical team and my family kept me well informed while I was an inpatient and I was confident that she was in

good hands which allowed me to focus on my recovery.

My surgery and recovery was a big deal even without any major complications. Coming off the epidural pain relief around day three was really rough, and it took a while for me to get back to full strength, but I have absolutely no regrets and I'd do it all again in a heartbeat.

ABO incompatible donation

Every person is born with a particular blood group (or blood type). The blood is classified according to the presence or absence of certain substances, called antigens, on the surface of the red blood cells. The most important blood group system is the ABO system. Individuals may have the A antigen (blood group A) or the B antigen (blood group B) on the surface of their red blood cells. Alternatively, they may have both antigens on the surface of their red blood cells (blood group AB) or they may have neither antigen (blood group O). See diagram.

Most children receiving a liver transplant will receive an organ (or part of an organ) from a donor who has a "compatible" blood group to their blood group. This means that the organ comes from a donor who does not carry a different antigen on the surface of their red blood cells than the recipient. The concern about transplanting an organ from a person whose blood cells carry a different antigen is that the immune system of the recipient will recognise that antigen as foreign and mount an attack against it. This can lead to rejection or complications affecting the bile ducts and blood vessels. At worst, the rejection can be very aggressive and may lead to loss of the new liver.

In practice, this means that children with blood group A can receive an organ from someone with blood group A. However, they can also receive an organ from someone with blood group O (because individuals with blood group O do not have any of these antigens on the surface of their red blood cells). Similarly, children of blood group B can receive an organ from

someone of either blood group B or blood group O.

Children with blood group O would only usually receive an organ from a donor of blood group O because the cells from donors of either blood group A or B will display antigens that are foreign to the recipient.

On the other hand, because children of the rare blood group AB carry both the A and the B antigens on their red blood cells, they can receive an organ from donors of blood group AB, but also from donors of blood groups A, B and O.

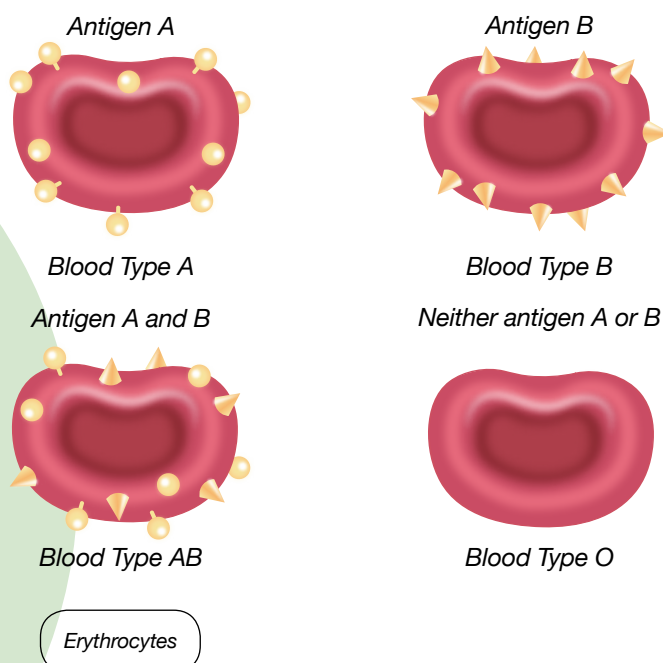
Although transplanting according to blood group compatibility reduces the risks of complications, it does limit the availability of organs to children on the waiting list.

In certain circumstances, the transplant team may recommend considering that your child receives a liver (or part of a liver) from a donor with an incompatible blood group. This might

be considered if your child is very sick while waiting for transplant and it is felt that the risks of waiting for an organ from a compatible donor are greater than the risks of using an organ from someone of a different blood type.

In recent years, a lot of research has been carried out to look at ways to get around the increased risks of rejection with transplantation across blood groups (ABO incompatible transplant). With newer and more powerful immunosuppressant agents, the risks of rejection in ABO incompatible transplantation have been significantly reduced and are now approaching the risks for ABO compatible transplants. However, the use of stronger immunosuppressant agents may be associated with an increased risk of infection.

The transplant team will discuss all of these issues with you in detail if they are considering an ABO incompatible transplant for your child.



First Steps: the transplant assessment

The major steps involved in having a transplant are:

- Assessment
- Waiting
- Getting a donor
- The transplant operation
- The first few weeks/months after transplant
- Life after transplant

What is the Assessment?

The assessment process aims to achieve many things and has to be thorough. The process involves:

- Evaluation of any issues and risk factors which might occur with respect to a transplant, including other health issues
- Determination of the urgency of the need for transplant
- Assessment of the diseased liver with respect to treatment while waiting for a transplant
- Determination of the size, blood group and type of donor liver required
- Consultation with other specialists if needed, and decide whether to place your child on the transplant waiting list.

Most importantly, the assessment process gives you time to meet the transplant team and have a better understanding of the transplant process, and gives you the opportunity to ask questions. The purpose is to provide you with sufficient information to enable you to give informed consent at the time of transplant.

Who is my contact person?

The Liver Nurse Specialist (CNS) will be your main link person during this process and will guide you through your assessment, ensuring that you understand all aspects of transplantation. We hope that it will be possible for both parents to be available during the assessment period. If this is difficult due to work commitments, the CNS will highlight which sessions are important for both parents to attend. We will be very happy to write letters to employers to help make it possible for both of you to be there. Alternatively we welcome other members of your family to attend the sessions to provide you with valuable added support.

If it is impossible for both parents to attend for the whole assessment process, the CNS can set up a time to call your home and provide information and answer any questions that you may have. (We currently have two CNS, whose contact details are in the phone list at the back of this manual).

How long will the Assessment take?

To some extent, this depends on the urgency of the transplant. In general, the assessment will take an average of five days, commencing on a Monday and finishing by Friday afternoon. However, for acute liver failure the process may need to be shortened and can be done in a day.

Your child will undergo a thorough evaluation that consists of a variety of

Blood tests and investigations to ensure that their other organs will be strong enough to carry them through the operation. You will also meet with various members of the Transplant Team. The assessment process is designed to give an overall picture of your child's health and to inform you fully about liver transplantation.

On the following page is a checklist of health professionals you will meet throughout the assessment period. You will have met many of these people before, but for others this will be the first point of contact.



Professional Seen	Name	Date
Hepatologist		
Transplant Surgeon		
Liver CNS		
Anaesthetist		
Social Worker		
Developmental team		
Dietitian		
Pharmacist		
Psychologist		
Renal (Kidney) Doctor		
Dentist		

Your Transplant Team

All members of this team work together, with a view to providing the best possible care for each child/young person. Each team member has a specific role to play in the assessment process. Some will be described in more detail later.

Once all the team have made their assessments, the information is put together and the team make the decision as to whether your child will be able to be put on the transplant waiting list. This decision will be reported back to you at the earliest opportunity by the Gastroenterologist/Hepatologist and CNS responsible for your child's care.

Where will I stay?

If you live within the greater Auckland region and transport is not a particular problem, you will be able to return home each day after the scheduled investigations.

If you need time out during the day between investigations but need to remain on site, you are welcome to use the Ronald McDonald Family Room facilities situated on level three of Starship Hospital. Any member of staff will be happy to point out the location.

If your child is clinically unwell, it may be necessary to admit him/her. The ward can provide room for one caregiver to

stay overnight with your child. (Please note the ward can not accommodate siblings overnight, unless you have a young breast feeding infant.)

For families who live outside the Auckland region, the NZLTU will fund travel to and from Auckland (limited to one child and one caregiver), and accommodation at Ronald McDonald House for the assessment period, whilst waiting for a liver transplant and for 3 months post-transplant. The CNS will arrange this for you. For visits to Starship outside the periods outlined above, your local hospital will make arrangements through the Ministry of Health (MOH). It may be possible for

the social worker in your local hospital to make an application to the MOH to fund a second caregiver to attend Starship for important visits.

Inpatient stays will usually be on Ward 26B. The team of health professionals working on 26B specialise in the care of children with complex and chronic health care needs. The nursing team is led by Sandra Murphy who works Monday to Friday. If you have any concerns relating to your child's ongoing nursing care, please approach Sandra so that issues are resolved in a timely manner.

Phone 09 307 4949 extn 25760.



Ronald McDonald House

Assessment Tests/Investigations

Depending on your child's illness a number of investigations will be undertaken. Each investigation or consultation will be explained. The CNS can be contacted to answer questions and guide you and your family through the week. Don't be afraid to ask questions.

The following is a brief list and explanation of some of the common investigations done during this time.

Not all of the following will apply to everyone:

Ascitic tap

A small amount of fluid is drained from the child's abdomen by inserting a needle through the skin. The fluid will be tested for any infection and protein levels will be measured. Larger amounts may be removed if the abdomen is too distended or swollen.

Biopsy

A liver biopsy is where a small sample of tissue is taken for examination. This is performed under general anaesthetic and guided by ultrasound scanning. After the biopsy it will be necessary for your child to remain in hospital overnight for monitoring.

Blood tests

Routine blood tests include: clotting, kidney function, liver function, blood group, viral studies (CMV, hepatitis, HIV), glucose and full blood count (anaemia, infection).

Cardiac echocardiogram ("Echo")

Similar to a liver ultrasound scan, except it scans the heart.

CT Scan

A computerized x-ray scan of parts of, or the whole body. A GA (general anaesthetic) may be needed in young

children, as they need to lie very still during the scan.

Chest X-ray

An x-ray of your heart and lungs.

Electrocardiograph (ECG)

Small electrodes are placed on your child's chest, arms and ankles and a recording of the electrical activity of their heart is taken. This is painless and will take only 5–10 minutes.

Endoscopy

Under GA, a tube is inserted through your child's mouth into the stomach. A tiny camera at the end will show any abnormalities in the oesophagus and stomach.

Chromium EDTA GFR (Glomerular Filtration Rate)

This test measures how well the kidneys are functioning. It is done by our nuclear medicine department. An IV (intravenous) line is placed in the arm of your child, and he or she is then given an injection of dye. Blood tests are then taken at 2, 3, and 4 hours after injection.

Lung function

The child/young person will be asked to blow into a tube attached to a measuring device, which records the amount of air exhaled. These tests are only attempted in children over the age of six years, as the ability of the child to understand and physically do the test is vital to the effectiveness of the study.

Magnetic Resonance Imaging

Imaging which uses powerful magnetic radio waves and a sophisticated computer to create detailed pictures of the body. It normally takes 30 to 60 minutes and is done under GA for young children.

PTC (Percutaneous Transhepatic Cholangiogram)

A needle is passed through the skin and into the liver guided by ultrasound. Dye is injected into the biliary duct which can then with the use of x-rays show any blockages and/or narrowings this requires a GA.

Ultrasound scan (USS)

A clear gel is applied to the abdomen and a handpiece (or transmitter) is moved across the child's abdomen which visualises blood vessels, bile ducts, the liver, pancreas, spleen and kidneys. Usually your child will need to fast for 4–6 hours before an ultrasound.

This scan is repeated every 8 weeks while waiting for transplant (or more frequently if there are specific concerns).

Vaccination History

Where possible all children on the waiting list are fully vaccinated using an accelerated schedule. All vaccinations will be recorded in both clinical notes and the Well Child Book, in addition a notification will be sent through to the National Immunisation Register'. Administration of any live vaccines during this time will mean the child or young person will be suspended from the active transplant list for a period of four weeks. This will be fully discussed with you as parents each time.

Roles Of Other Specialities

Social Worker

Our Social Worker will see you as part of the assessment process and will discuss with you your social and family history. He/she is available to discuss with you any aspect of your hospitalisation which may cause you or your family concern. They can arrange counselling and can provide information or practical support, depending on your needs and situation. Our social worker can also assist you with liaison with other agencies regarding any financial or housing worries you might have.

Liaison Psychiatrists

Families living with chronic and life threatening illness often struggle with the emotions generated by these stressful circumstances. One of our Consult Liaison psychiatric team will meet with you and your child/young person during assessment. This service is here to help you through this time and can help to find ways for you and your family to cope with the emotions that this stressful time can generate.

It is possible to self refer by talking with the CNS, who will arrange a consult for you. Alternatively a member of the transplant team may approach you and ask if you would like to meet the Consult liaison team. Your consent is required for any referral to be actioned. Referrals to this service can be made during any other period of hospitalization, or even when you are at home if you feel this service is needed.

Of note: Where possible the consult liaison team will meet with you in a urgent or 'crisis' situation. However in most situations the visit

will be scheduled as an outpatient appointment.

Renal Physician

As part of the assessment process, your child will have a number of investigations to assess their 'base-line' kidney (renal) function. Our kidneys do the following to keep us fit and healthy:

- Regulate the composition of your blood
- Keep the concentrations of various ions and other important substances constant
- Keep the volume of water in your body constant
- Remove wastes from your body (urea, ammonia, drugs, toxic substances)
- Keep the acid/base concentration of your blood constant
- Help regulate your blood pressure
- Stimulate the production of red blood cells
- Maintain your body's calcium levels

We have come to understand that, over time, immunosuppressant medications can affect how well the kidneys are able to function. We work closely with the renal service to detect and prevent damage to the kidneys.

Dentist

Chronic liver diseases often causes bad teeth, so good dental care is part of the transplant process.

Maori Health Services: Te Whanau Atawhai

A service available to all patients and their whanau/family. They provide a valuable link within the team. As a team we believe early recognition of family structure, supports and cultural needs is imperative to creating positive working relationships as we work together to keep your child as well as they can be. The cultural support team are available to help you and will

advocate for the needs of you, your family and your child, and will assist in ensuring mutual understanding.

Chaplains

A chaplaincy service is available. This can be used as a support through your assessment and also after your transplant, for both you and your family.

Pacific Island Support

A service available to people of pacific origin, supporting families in the hospital setting.

Play Specialist

Play is the work of childhood, which ensures development continues normally. Prolonged hospitalization significantly reduces a child's opportunities for exploration and play. Our play specialist team mainly come from an early childhood education background and where able will provide opportunities for your child to play, or provide toys that will stimulate the senses in infancy. We are fortunate in Starship to also have access to an adolescent play specialist who will provide youth friendly activities and programs.

A big component of the play specialist role is in preparation of children for procedures, 'procedural play'. They can also help your child to express their fears and anxieties associated with hospitalization in a safe and non-threatening environment.

Most children who require a liver transplant are infants. However, for older children/young people arrangements will be made for them to spend time with a member from the play specialist team.

The Waiting List

Waiting is hard on everybody, particularly if your child is getting sicker. However, your team will do everything they can to keep your child in as good a shape as possible, for as long as possible, and rest assured, the system for finding a donor does work! We would advise you to try to live as normal a life as possible.

How long will our child have to wait?

There is no answer to this. It all depends on finding the right liver for your child. This depends on blood group, size matching and other factors. It may take anything from one day to more than one year. The average waiting time is 4–8 months, unless a suitable living donor is available, at which time a transplant can be scheduled for a particular date.

How are deceased donor livers allocated?

There are three main considerations

- Medical Urgency (justice/equity)—sickest is transplanted first
- Utility—the best use of the transplanted liver (? Is the liver splittable how can they benefit the most people?)
- Transplant benefit.

The New Zealand Liver Transplant surgical team factors in all three considerations when making their choice when allocating a donated liver. The first factor is the most important. The other two factors are considered when there are a number of possible recipients who are equally unwell, this helps the team to come to the best decision when faced with difficult circumstances.

Where will our family wait?

If you live within 5 hours travelling time (with good access to regular flights) of Starship and your child remains stable, you may wait at home. Unfortunately if you live more than 5 hours travelling time away, you may have to wait in Auckland when your child is near the top of the waiting list for his/her blood group, or if the transplant team feel your child is too unwell. Once again you will be accommodated at Ronald McDonald House.

Who will let me know when a liver becomes available?

Most families can be contacted by the liver transplant coordinator on their mobile phone. However, if this is not an option we will provide you with a pager through which you can be contacted. You may be contacted any time night or day.

Please keep your phone in credit and charged. In the case of a pager, change the battery monthly. It is important that you carry the phone/pager with you at all times. If we cannot contact you quickly, your child may miss an opportunity to receive a suitable liver and continue waiting.

What must I do when I get a call?

Keep calm! Ensure your child has nothing further to eat and drink. Travel plans will already have been prearranged and discussed with the CNS. Carry out these plans as arranged. A bed on ward 26B will be ready and waiting.

Have your bags packed! If you are from outside Auckland please have a pre-packed bag for each family member, to aid a well-timed departure, as it may

be some time before you will be able to return home. This will help to reduce stress levels in an already stressful time (especially if a sibling's precious bear/toy is accidentally left behind.)

Ensure you pack:

- Your child's usual medications, favourite toy and a couple of other familiar and loved possessions.

Other things to do while you wait:

- Spend quality time together as a family, talk to your child and their siblings about what is happening. Honest communication will not remove their anxieties but may prevent them becoming overwhelming and too scary. Let the CNS know if your child is unwell. The transplant team must be aware of your child's condition immediately if they become unwell.
- Ensure all your other children's vaccinations are up to date. Check your pet's vaccinations are up to date – and ensure you know who will care for them in your absence.

Where do I report to when I arrive in hospital?

You should go to Ward 26B which is on the 6th floor of Starship. The nursing team will be expecting you, and have allocated an appropriate room prior to your arrival.

What will happen when I arrive?

There will be lots to be done upon your arrival at the hospital. You will meet with the doctors and other team members who will assess your child's 'fitness for theatre'. The on-call liver transplant coordinator will be keeping the team updated with any changes, and where possible will provide you with an expected Theatre time.

The following investigations will be necessary:

- Chest X-ray
- ECG
- Blood tests

You will need to ensure your child has a shower or bath.

The doctor will ask you to sign a consent form.

Will my child be the only potential transplant recipient called into the hospital?

Sometimes two potential recipients are called into hospital. The second person is the “back-up”. (You will always be told if your child is being called in as the “back-up” recipient). This means that if the person at the top of the waiting list is found to be unsuitable (before or during the operation), then the “backup” person can be transplanted immediately without wasting the donor liver. Only certain transplant candidates need a “back-up”. The person who receives the liver will always be the most suitable recipient.

Occasionally, the donor liver may be found to be unhealthy once examined by the donor surgeon and will be deemed unsuitable. If this occurs, your child’s transplant will be cancelled. However if for any reason the operation does not take place, your child will automatically go back to the same place on the waiting list until another liver becomes available. This can be very disappointing, but it is better to wait a little bit longer and get a good match.

The Waiting Period

We acknowledge that this is a period of great uncertainty and anxiety for a family. As a team we aim to minimise the disruption to ‘normal family life’ by attempting to keep your child as well as possible while they await their transplant.

We would advise you to try and live as normal a life as possible. If your child is school aged, allow them to attend school part time if they remain well enough.

Attempt to stay connected with your family and friends, and where possible take time to enjoy some of the simple pleasures. It’s ok to have a laugh and to take some time out for yourselves as parents.

If your friends and family have infectious illnesses, suggest that you would prefer a phone call while they are unwell and look forward to a visit when they are once again feeling better.

Let the CNS know if your child is unwell. The transplant team must be aware of your child’s condition immediately if they become unwell.

If your child is in hospital for an extended period

- Take the opportunity to access the hospital grandma service—to allow you some time out.
- Let your friends and family know what is and is not helpful—practical things like pre-prepared meals can be helpful—or a delivery of reading material and chocolate!
- Spend quality time with your other children. Talk with them about their brother or sister’s illness, allow them to ask questions—also ensure you have time together that is just about them—have fun together.

While your child awaits Liver Transplantation the team will focus on optimising nutrition and minimising complications associated with end stage liver disease. This may mean your child requires regular infusions of Albumin, antibiotics and doses of Vitamin K. The team may also discuss with you the placement of portacath (port). This is a form of central venous access. We will discuss the risks and benefits with you more fully should it become necessary.

Complications of chronic liver disease

The complications of chronic liver disease in children include:

- Malnutrition and faltering growth
- Portal Hypertension
 - ◊ Varices
 - ◊ Ascites
 - ◊ Peritonitis
 - ◊ Hypersplenism
- Hepatocellular Carcinoma
- Encephalopathy
- Coagulopathy

The following paragraphs explain these complications.

Malnutrition and Faltering Growth

Malnutrition in children with liver disease is due to a combination of poor intake because they are unwell, poor absorption because of the lack of bile getting to the intestine, poor delivery of nutrients to the body, and increased nutrient requirements because of the liver's metabolic problems.

Our aim is to achieve optimal (best) nutrition for your child. This will involve, frequent monitoring by the dietitian and medical/nursing team. Maintenance of adequate growth is an important part of the treatment of your child's liver disease. They will not be able to absorb all the nutrition from their food and will need extra help. Special formulas and vitamin supplements are necessary to ensure your child has sufficient nutrients for growth. Additional measurements, to assess their body fat and protein stores, will also be taken.

Your dietitian will develop a practical plan with you to ensure your child receives sufficient nutrition to achieve as close as possible to their growth potential.

Infants with Liver Disease

If your child is not growing adequately, with breast feeding alone, they will be started on a formula containing special fats and hydrolysed (broken down) protein designed to be utilized more easily by their body. The concentration can be increased to supply additional nutrients and promote weight gain and an energy powder or liquid can be added. These products are available on prescription with a PHARMAC subsidy and your dietitian will give you a special recipe. If your child is consistently unable to take sufficient volume of formula, from their bottle, a nasogastric tube will be used to "top up" their feeds with formula. A pump can be used to "continuously" tube feed a child unable to tolerate large amounts of feed at one time. Your dietitian will give you a feeding plan.

It is important that your baby is offered solids when they are ready. You can discuss this with your dietitian. Small amounts of solids are important at meal times to develop feeding and language skills but their formula is the most important source of nutrition.



Aria, who had liver and intestinal failure



Lara, Isabelle, Jaya and Hamish with their naso-gastric feeding tubes

Older Children and Adolescents with liver disease

Older children and adolescents will be encouraged to eat high energy meals and snacks to ensure that their growth is as good as possible and their full height potential is achieved. Special supplementary drinks may be required. Sometimes a NG (nasogastric tube) is needed for additional nutrition.

It is important to eat regularly and aim to have three meals a day with snacks in between. This may require some planning if a family meal is missed eg have a meal left in the fridge. Sometimes a special diet will be necessary and your dietitian will give you advice on this.



“Isabelle was fed predominantly via an NG tube for seven months pre-transplant and a further month following transplant.

It was stressful and messy at times with frequent vomits, the tube being pulled out, the end cap coming off accidentally and draining into the cot or on the floor, and more. But it was a huge relief to watch Isabelle go from skinny arms and legs and big tummy – transformed to a chubby baby with plenty to ‘come and go on’ going into transplant time.

This was during her first year and her introduction to solid food was pretty unsuccessful, and I worried about her learning to eat and her speech development. But after transplant her appetite came back quickly, she started eating and drinking without much hassle and within a few weeks the tube was gone. ”

– Jodie, mother of Isabelle.

The difference maximising nutrition makes



The babies show the difference in growth between twin boys at age six months - one of whom had liver disease



The same twin boys at ten months of age after maximising nutrition

Portal Hypertension

This is an important complication of liver disease which is closely monitored pre-transplant. Portal hypertension is a common problem in children with liver disease. As the liver becomes increasingly scarred, it gets stiffer and it is difficult for blood to flow through it. Blood has to make its way back to the heart through other ways. Increasing amounts of blood have to flow through veins that are not designed to cope with that.

What normally happens?

The heart pumps blood to the intestine where the nutrients from food are picked up. The energy laden blood is then pumped to the liver for processing. The blood then drains back to the heart.

What is portal hypertension?

Portal = the blood vessels that drain the stomach and intestine

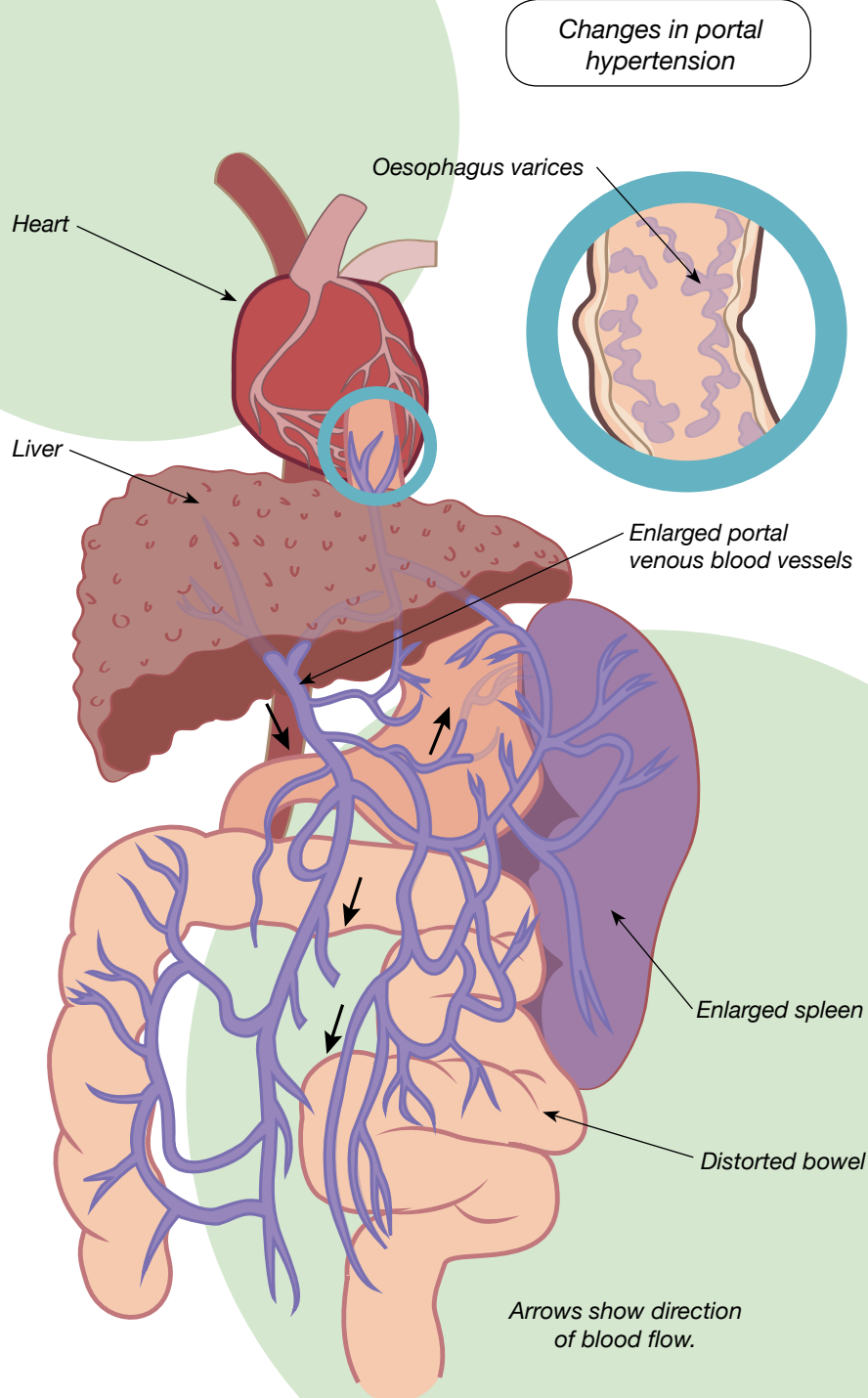
Hyper = high

Tension = pressure

Portal hypertension is high blood pressure in the blood vessels draining the stomach and intestine.

Why do patients with liver disease have portal hypertension?

In liver disease the scarring in the liver makes it hard for blood to pass through it. The blood flows back to the heart via different paths. It is this increase in blood flow through alternative routes that is called portal hypertension.



How will my doctor know if my child has portal hypertension?

- If the spleen is larger than normal, this suggests portal hypertension. The spleen size can be measured by the doctor examining your child's abdomen or by ultrasound scan.
- Blood tests: A low platelet count or white cell count may suggest portal hypertension.

What problems does portal hypertension cause?

There are three major problems that can happen if you have portal hypertension.

- Varices (varicose veins of the intestine)
- Increased spleen size and function
- Ascites (swelling of the intestine and fluid leaking out)

Varices

Varices are "varicose veins" of the intestine. When blood finds it difficult to go through the liver, it makes it way back to the heart through alternative blood vessels. These blood vessels are not always able to cope with the increase in blood flow. The increase in pressure in the veins makes them larger and some of them have thin walls. This leads to a risk of bursting which may cause a serious, or even life-threatening, bleed.

Where can bleeding occur?

The common places that bleeding can occur from are from the oesophagus (food swallowing tube), stomach, and from the rectum (bottom).

How will I know if my child has a bleed?

1) Vomiting blood

Blood in the vomit may be fresh (bright red) or altered (clots or old, black blood)

2) Passing blood in their stool

Blood in the stool is usually old and causes the stool to be black, tarry and strong-smelling. However, fresh (bright red) blood may also be passed. Note: this is different from the common symptom of flecks of bright red blood around stools that might occur from constipation.

3) Becoming very pale or fainting

What do I do if my child bleeds?

This can be life-threatening and requires immediate medical attention.

What can be done to prevent bleeding?

If your child has signs of portal hypertension (a large spleen or low platelets) an endoscopy may be performed. Your child may also be put on medicine to make it less likely that they will bleed.

Your child will be stabilised in the local hospital and transferred to Starship as soon as possible.

**CALL 111 FOR AN
AMBULANCE
(No matter the time
day or night).**

Tell the Ambulance Operator (Com's) what has happened and that your child has portal hypertension and liver disease.

Take anything that might help confirm that there was bleeding (bowl, blood stained clothing etc)

At the hospital your child:

- Will have a line (cannula) sited and will have blood tests
- May need special medication into the vein to decrease bleeding
- May need an endoscopy

Ascites

What is ascites?

Ascites is extra fluid inside the abdomen, but outside of the intestine, created when the liver is unable to make enough proteins.

How do I know if my child has ascites?

If your child's abdomen is getting larger – particularly if this happens rapidly (over days to weeks) then they may have ascites. Your doctor will be able to tell by feeling your child's abdomen or by an ultrasound scan.

How is ascites treated?

Ascites is usually treated with medication and in the setting of end stage liver disease with a combination of medications and albumen infusions.

Can ascites get infected?

Yes - the fluid can get infected, this is called peritonitis, if your child has an episode of peritonitis while awaiting transplant they will be suspended from the list until the infection has resolved.

Peritonitis

How will I know if my child has peritonitis?

If your child displays the following symptoms please bring him/her to the hospital immediately.

The usual signs of peritonitis are:

- A rapid increase in the size of their abdomen.
- Pain in the abdomen.
- Fever.
- Diarrhoea or vomiting.

Peritonitis is treated with IV antibiotics.

Summary

Portal hypertension is an important problem in liver disease.

Things to look out for are:

- 1. Bleeding - If your child ever vomits blood; fresh blood – red, old blood- black and sticky or passes stool which is fresh (red) or old (black). They are likely to be bleeding. Your child needs to go STRAIGHT to HOSPITAL.**
- 2. Abdomen getting larger. If your child's abdomen becomes suddenly larger particularly if they are unwell then your child needs to be seen by a doctor that day.**

Hypersplenism (overactive spleen)

What is the spleen?

It is an organ that usually is found on the left hand side of the abdomen, behind the stomach.

It has two important jobs:

1. New blood cells are always being made by the bone marrow, the spleen has the job of getting rid of the old, tired ones
2. It is an important part of the immune system (bug fighting).

What happens in liver disease?

The increased blood flow through the spleen makes it grow larger.

Why is a large spleen a problem?

There are two major problems with having a large spleen:

1. A large spleen becomes too efficient at getting rid of blood cells. When it is enlarged it can get rid of cells which are still new and useful. The commonest problem is a decrease in platelets (clotting cells) and white cells (bug fighting cells). It is uncommon for red blood cells (oxygen carrying cells) to be decreased.
2. The spleen is delicate and can be damaged if rough contact is made, more so when it is enlarged. If damaged it can cause bleeding into the abdomen.



Oliver pre and post transplant, celebrating his first birthday 2012



Hepatic Encephalopathy

Is a sign of end stage liver disease, characterised initially by confusion, increased irritability, altered sleep patterns, and in advanced cases altered levels of consciousness and coma due to brain swelling.

In the mildest form it can be very difficult to detect, in a school aged child you may observe worsening school performance and poor concentration and forgetfulness.

It is very difficult to accurately detect encephalopathy in infants, due to their inability to communicate. Parent observation of changes in their child is key to making a diagnosis of encephalopathy.

Things to look out for:

- changes in sleep routines
- increased irritability and
- difficult to console
- a high pitched cry

Coagulopathy

Is a failure of the liver to make clotting factors leading to increased risk of prolonged or excessive bleeding. This sometimes occurs due to a lack of vitamin K but may also be a sign that the liver is failing to synthesise (make) clotting factors.

It is vital that your child receive all prescribed doses of Vitamin K, it may be necessary to give them additional

Intravenous (IV) doses of vitamin K if the oral solution is not sufficient to control your child's INR within normal range.

Things to look out for:

- Unexplained bruising
- Prolonged bleeding
- Unexplained nosebleeds

Hepato-pulmonary syndrome

Can occur in a minority of children with advanced liver disease. (both acute and chronic)

What is Hepato-pulmonary syndrome?

It is a problem of gas exchange in the lungs, which can lead to low levels of oxygen in the blood stream (hypoxia) and respiratory failure. It is reversible within the first few weeks post transplant in most children.

Things to look out for:

Pre transplant close observation of your child's skin colour, work of breathing and tolerance of exercise is important. From time to time we will check your child's oxygen saturations in the clinic or ward setting and arrange for more advance testing if we are at all concerned.

Children with Hepato-pulmonary syndrome will most likely require oxygen support when recovering from any episode of acute illness.

Hepato-renal syndrome

What is Hepato-renal syndrome?

Hepato-renal syndrome is a late complication of advanced liver disease, and portal hypertension. It is a progressive worsening of kidney function which can result in kidney failure.

It can be caused by significant fluid shifts within the body which can reduce blood flow to the kidneys. Possible causes are dehydration, infection and blood loss.

Things to look out for:

In the mildest form there will be changes in the bloods (low sodium) and difficulty to effectively pass accumulated fluids within the body.

In the moderate form there will be changes in appetite, and reduced energy levels.

In late stages, the child will have ongoing nausea, vomiting and excessive thirst. The child may be drowsy.

It is often difficult to identify if these symptoms are caused by Hepato-renal syndrome or another complication of liver disease which have similar symptoms, but this will be clarified by blood tests and further investigations if necessary.

The Operation

When you get 'the call', don't panic! Everything is under control. It is often a relief to know the day has finally arrived. It will be a long day, but you'll get through it ok. After the operation you can expect a few critical, stressful days in the PICU, then a gradual recovery on the ward (26B).

Where do I report to when I arrive in hospital?

You will need to report to Ward 26B, which is on level 6 of Starship. The nursing team will be expecting you, and will have allocated an appropriate room prior to your arrival.

How is the operation carried out?

The surgeon will make an incision in your child's upper abdomen. This is called a 'Mercedes Benz' incision. The old liver is removed, first by cutting the ligaments that hold it in the abdominal cavity, and then by clamping all the major blood vessels to prevent bleeding.

The new/donor liver is inserted and the blood vessels connected starting with the hepatic vein, then the artery, then the portal vein. After this, blood is allowed to flow through the new liver by removing the clamps from the major vessels.

When the new liver begins to receive blood again the connections are tested. We usually know if the liver is working by this stage because it starts to produce bile fairly quickly.

The bile ducts will either be connected

to a loop of intestine called a Roux-en-Y or Roux loop (see labelling of 'Anatomy in paediatric liver transplant'), or in older children stitched directly to the patient's own bile ducts. If your child has biliary atresia they will already have a Roux-en-Y from their Kasai operation and this will be re-used.

The abdomen will be sewn using stitches which dissolve and which do not need to be removed. Post-operatively you can expect dressings to be replaced at least every 3 days and more often if necessary.

Your child can receive either a whole liver, or more commonly a split or reduced size graft.



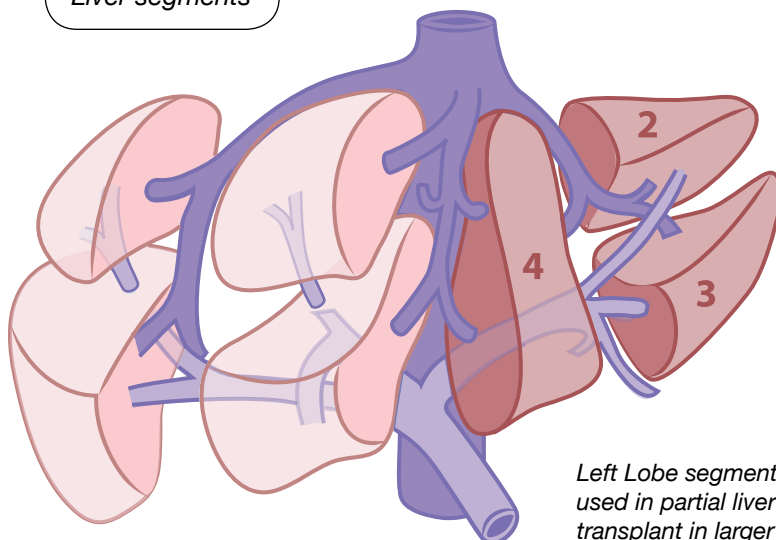
5 weeks post transplant



22 months post transplant

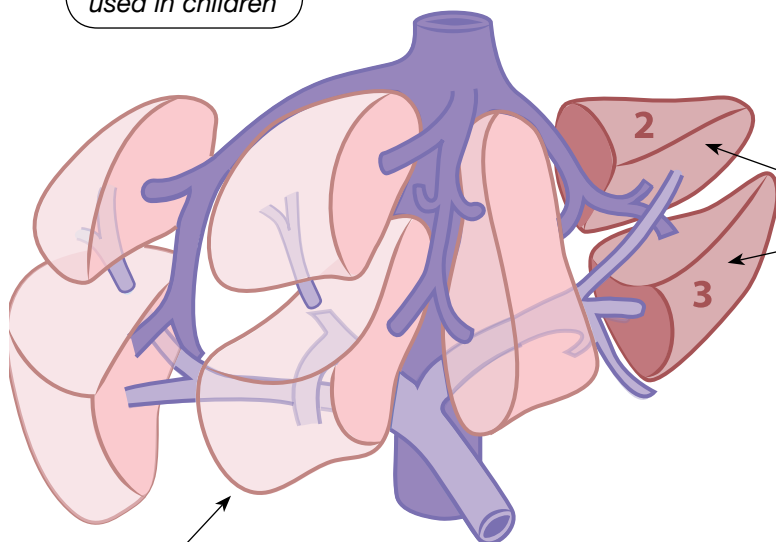
New Zealand and Australia share donor organs and, if a split transplant is undertaken, the remaining segments may be sent on to an Australian centre or vice versa.

Liver segments



Left Lobe segments used in partial liver transplant in larger children.

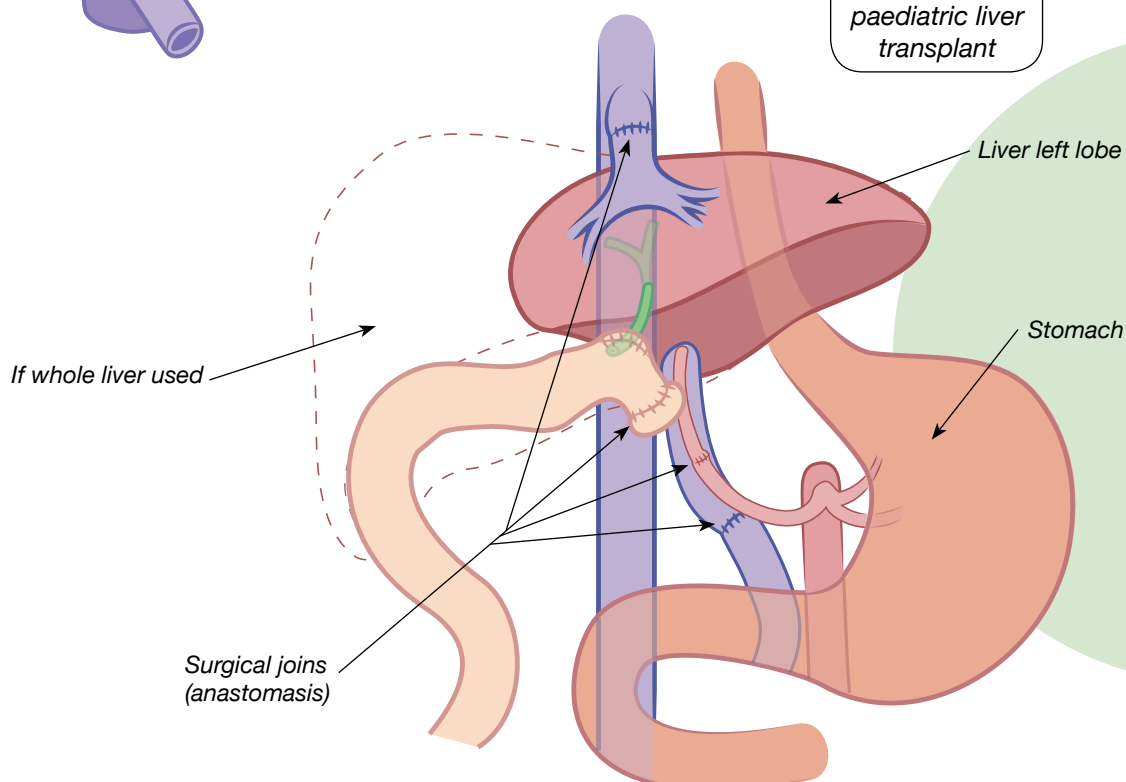
Liver segments used in children



Right lobes

Segments used in smaller children and babies.

Anatomy in paediatric liver transplant



Braxton day 1 post liver transplant



What happens immediately after the operation?

Immediately after the operation your child will be taken to the Paediatric Intensive Care Unit (PICU), where they will usually remain for 24- 48 hours, or longer if necessary. Care in PICU is shared between the transplant service and the intensive care team. The transplant team will do a ward round twice daily around 8-9 am and 4 pm. Your child will also be reviewed twice daily by the Starship pain service to ensure that they are as comfortable as possible.

There are lots of tubes! Initially a breathing tube connected to a ventilator will help your child to breathe effectively and clear secretions until they are strong enough to do this for themselves. Although the tube is not painful, it can be uncomfortable. Once the tube is removed it will be replaced with an oxygen mask.

Your child will have up to two wound drains on each side of their abdominal wound. These will remain in place to allow fluid collecting in the abdomen to drain out until drainage is minimal. Your child will also have 2 central lines (even if he/she has a port), an arterial line for blood pressure monitoring and a urinary catheter. It is important to measure your child's blood pressure and urine output every hour for the first couple of days.

Immunosuppression medications are started, and antibiotics to prevent infection are administered. We closely monitor everything. There are three

specific early complications which we look out for:

1. Surgical problems such as bleeding
2. An infection
3. A problem with the donor liver, such as it not working well or a problem with blood vessel connection – see the section on 'Complications after transplant' for details.

Parents and caregivers have 24 hour access while their child is in the

PICU. Please be aware that due to safety considerations no sleeping accommodation is provided for caregivers - you are welcome to use the family lounge or the veranda area to relax. Where possible arrangements will be made for you to stay in one of the Ronald MacDonald family rooms located on level 3 of Starship.



Baby Oliver surrounded by medication pumps and monitoring machines in PICU

The first few weeks/months after transplant

A lot happens in the first few weeks/months following the transplant. This is a time of recovery, close monitoring in hospital, dealing with early complications if they occur, getting established on immunosuppression (drugs to prevent rejection) and rehabilitation.

What happens in the ward?

When well enough, your child will be transferred from PICU to Ward 26B under the care of the gastroenterology and transplant surgery team.

Daily ultrasound scans are carried out to visualise the blood vessels, bile ducts, liver and spleen for the first 5 – 7 days post transplant. In addition, if your child's liver tests become abnormal he/she may require a liver biopsy, looking for signs of rejection. Because the new liver does not have nerve connections on its surface the biopsy procedure is usually painless but a general anaesthetic is used.

Because the nerves have been cut during the operation he/she may experience a sensation of numbness around their scar for many months following the transplant.

The catheters and drains will usually be removed within the first couple of days after the transplant. A nasogastric tube will be used for feeding immediately after the operation but will be removed once your child is able to eat properly. Your dietitian, after discussion with the transplant surgeons and gastroenterologists, will make a plan for feeding. Children who were nasogastrically fed pre-transplant will continue on their pre-transplant formula in the short-term.

Your child will receive regular visits from the physiotherapist who may administer chest physiotherapy to help

coughing and clearance of secretions. Alongside nursing staff, they will also assist early mobilisation of children old enough to walk.

What medications will your child be on?

Initially your child will be on a wide range of medicines including anti-rejection drugs (immunosuppression), anti-fungals, anti-virals, antibiotics and medicines that lower blood pressure (see sections on 'what are immunosuppression drugs' and 'Medications').

The medicines each play a very important part in preventing both rejection and infection, and combatting side effects of other medicines. It is important to report anything that you observe which might be a side effect to the transplant team.

What complications might be expected?

Some sort of complication is likely to occur, as it is uncommon for a transplant to go absolutely without incident, but you should know that these are mostly manageable (see section on 'Complications after Liver Transplant'). Rejection needs extra treatment in about 40% of children within the first month or so, we monitor and maximize the doses of anti rejection drugs during this time, gradually reducing them after that.

Because the immune system is suppressed, about 50% of children get some sort of infection during the first 3 months, varying from simple wound infections and viruses to more serious infections such as infection in the bile duct (cholangitis).

Plumbing problems with one or other of the connections may occur. These sometimes require a procedure to revise

that connection. The most common plumbing problem relates to the bile duct, which can be both diagnosed and treated without surgery, by inserting a tube called a stent through the skin into the liver under a general anaesthetic called a PTC (or percutaneous transhepatic cholangiogram).

Before going home

There is light at the end of the tunnel! Unless there are complications, most children can be discharged home 2-4 weeks after their transplant.

Before going home it is important to understand and get to know all the medications that your child is on (see section 'Medicines'). Usually there are at least four medicines (Prednisone, Tacrolimus, Cotrimoxazole and Valgancyclovir). Your CNS will take you through these in detail.

In addition, it is important that everybody at home understands and develops a hygienic lifestyle, being careful with hand washing, bathing, food handling, cooking and other activities (see section 'Life after Transplant: staying healthy').

What happens when we go home?

You will be followed initially twice a week with blood tests and clinic visits, gradually reducing to weekly, then monthly, then even 3 monthly as things settle down. By this stage the risks of complications are getting less and less, but episodes of suspected rejection (if they occur) will require a liver biopsy, and extra steroid treatment for 3 days. Infections are treated on their merits. Late bile duct plumbing issues might occur as the bile duct connections heal (see 'Complications after Liver Transplant' for further information).

Living with transplant



Life after Transplant

The aim of liver transplantation is to allow your child to have a good, rich and fulfilling quality of life.

The first few weeks and months after transplantation are challenging but as time goes on, the blood tests and visits to hospital become less frequent, the number of medications reduce and life begins to feel more normal again.

All the normal things that happen to every other child and family will happen to you. Indeed, the only abnormal thing will be that your child will remain on some form of immunosuppression and will require occasional clinic visits.

Life from a medical point of view is then a balance between the ongoing risks and side effects of immunosuppression and the relatively low risk of rejection. These risks are minimised by;

1. Leading a healthy lifestyle
2. Religiously administering the immunosuppression medications and getting timely lab and drug monitoring levels
3. Remaining vigilant about infection and infection exposure

Infection and rejection

Within 6-12 months, things like the risk of rejection episodes are likely to reduce to <1 episode per year, and we usually relax immunosuppression after about a year by reducing and stopping the prednisone, and allowing lower levels of tacrolimus.

However, while your child remains on any anti-rejection medication, you still have to be vigilant for the possibility

of infection or potential contact with infectious disease.

Bacterial infections are not common after the first 3-6 months, but virus infections are a particular problem in children on long-term immunosuppression. We monitor for several important viral infections (EBV, CMV – see section on ‘Complications following Liver Transplant’) using lab tests. We also want to know if there has been any exposure to certain viruses such as influenza, chicken pox, measles or glandular fever.

We ask to be informed about fevers, rashes, vomiting, diarrhoea, infection contacts. We recommend that all transplanted children and their families receive a ‘flu vaccine every year. The ‘flu vaccine is available in early autumn. The ‘flu is constantly changing and so the vaccine is different each year. This is why it is important to be vaccinated EVERY year. Please note the ‘flu vaccine is fully funded for the transplanted child and may be funded for other members of your family – please ask your family doctor for more details on this.

Bathing & hygiene

We recommend that your child is taught to be particular about their hygiene. Your child can take baths and showers after discharge from hospital with gentle washing around the wound area. Some children may have a bile drain. In this case, it will be necessary to try and keep this dry during bathing and showering. We also recommend your child learns to wash their hands frequently especially after using the toilet and prior to eating.



Skin care

People who have had transplants are much more prone to skin cancers. This risk is as high as 25 times more than in people without transplants. The risk is especially high in New Zealand where the sun is intense. Your child should not sunbathe and should not go out into the sunlight without high factor sun cream. Please look for sun creams with a high SPF (Sun Protection Factor) number. Avoid going outside for any length of time in the middle of the day during summer. However, please don't stop your child going out at these times – just be sensible.

Always make sure your child wears a wide-brimmed hat outside. If it is a very hot day, then long trousers and light long sleeves should be worn. Children should always have sun cream applied during the summer, even if it seems cloudy or overcast. Please remember to apply sun cream to areas of the skin often forgotten – for example, the tips of the ears, back of the neck etc.

Going out

Many families feel anxious about taking their children out after their transplants, particularly because of the infection risk. However, your child will spend many weeks and months recovering from their transplant and a gradual re-introduction to normal life is needed. In fact, trips out are more likely to make your child feel “normal” again, even if it is just to the dairy.

Avoid going out to very crowded enclosed spaces in the first few weeks and months e.g. cinemas, cafes, supermarkets, buses etc. Avoid people

with runny noses and coughs. There is nothing wrong with taking your child to these places during quieter times e.g. during the middle of the day on a school day.

Young children tend to carry more viruses and infections and are also more likely to catch these infections. Avoid large groups of young children initially e.g. children's birthday parties. Older children and adults tend to be less infectious and are less prone to infections as they have built up some immunity.

Do encourage relatives and friends to visit your child in small groups – this is an important part of the recovery process. However, please tell relatives and friends to stay away if they are ill or think they may be “coming down” with something. This may seem a little upsetting for them, but they will understand the need to be extra careful to protect your child.

School

We like our liver transplant patients to feel as normal as possible. It is important to try and keep up with their educational needs despite liver transplantation. After the transplant and as soon as your child feels able, a teacher from the hospital school will visit and start some gentle school work.

After discharge from hospital, it will be possible for schoolwork to be sent to your child at home. Obviously they will have less energy and tire more quickly for some time after the operation and so will not be up to a whole day of school work, but little and often is a sensible approach. Usually by 3

months post transplant, your child will be ready to go back to school, often part-time to begin with and building up to full-time. This is something we will discuss with you at clinic. Parents often feel anxious when their child returns to school, in particular about the risk of infection. It will be important for the school to know about the liver transplant and also to let you know as soon as possible if a classmate becomes ill with an infectious disease such as chicken pox. We are happy to write any letters of support to the school if needed. (see a copy of a school letter section in the appendix)

Kindergarten (kindy)

Pre-school children may be able to go to kindy from 6-12 months after transplant. This is something we will discuss with you at clinic. Often parents feel anxious when their child starts at kindy, especially about the risk of infection from other children. However, it is important for your child to have the opportunity to socialise with other children of their own age.

As above, it will be important for the kindy to know about the liver transplant and to contact you as soon as possible should any other child become unwell with an infectious disease. The disease we worry about most is chicken pox which is particularly common in pre-

school age children. Again, we will happily provide any letters of support that may be needed.

Sports and exercise

Exercise is important for everyone in order to keep as fit and healthy as possible and to lower the long-term risks of health problems such as high blood pressure and heart disease. Some of the drugs that are given after a liver transplant make high blood pressure and heart disease more common.

The risk of these problems is not high during childhood. However, when you consider that your child will be taking some of these drugs for life, it becomes important to stay as fit as possible. Another reason to exercise is to improve the tone/strength of the abdominal (tummy) muscles which are often weak because the abdomen has been distended before transplant and from the surgery.

You may notice that your child's tummy remains quite large after transplant. Some parents worry that this is because the liver and spleen are large or that there is fluid in the tummy. This is usually not the case and is usually just because of poor muscle tone. This will improve with time and exercise.



Brady preparing to surf*Sophie conquering the rock climbing wall*

We recommend aerobic exercise (running around type exercise, to make your heart beat faster and make you sweat!) for 30 minutes, 3-4 times per week as a minimum. Obviously this will need to be gradually introduced as your child will feel tired and weak for the first few weeks following their transplant.

Good sports include soccer, netball, dancing, swimming etc, but mowing the lawn would also do (not on a ride-on mower!) Contact sports which may injure the abdomen e.g. boxing, rugby

*Matthew preparing for the transplant games*

and some of the martial arts are best avoided for the first year or so after transplant and those children who have a big spleen may need to avoid them for longer. If letters are required for any sports clubs, we will be happy to provide them.

Obesity

Unfortunately obesity is becoming more common in people all over the World. This is partly because diets have changed and fast food has become more popular and also because we now take less exercise than we used to.

Importantly, people who are obese often accumulate fat inside their liver and this can lead to damage and scarring within the liver. Therefore it is important to try and keep your child fit and to avoid obesity. In addition, some of the drugs that are used after transplantation may increase the level of cholesterol and fat in the blood. For this reason, we recommend a healthy diet and plenty of exercise.

Your child may have had trouble gaining weight before transplant and

may have been on a high calorie diet or supplemental feed. With a normal liver, this is no longer necessary.

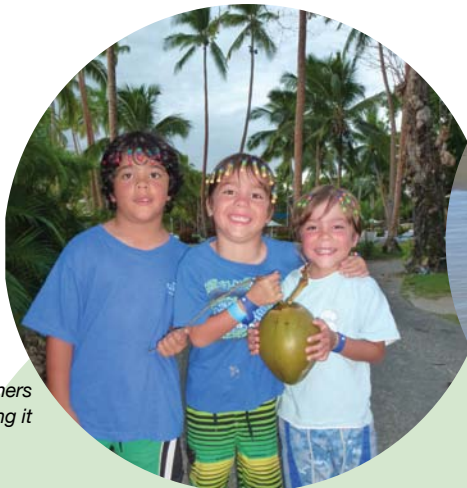
Another thing that happens to sick children is that caring relatives will often bring them treats such as lollies and fizzy drinks. We know that your child has been very sick and had a major operation and it can be difficult to refuse them "treats". As with many things these treats are fine in moderation but we recommend that they do not become too frequent.

Dental hygiene and care

There are several reasons why liver transplant children need to take special care of their teeth. Firstly, you may notice that if your child has been jaundiced as a young child that the first or "baby" teeth are discoloured and may look a little green. This is the jaundice staining the teeth. This is less common in the second set or "adult" teeth but if this occurs can be dealt with later by a dentist.

Secondly, your child may be receiving drugs e.g. cyclosporin that make the gums enlarge. In this case, teeth brushing is more difficult but even more important. We recommend that all our patients be encouraged to brush their teeth at least twice a day and that they are seen by a dentist once or twice a year for a dental check.

Please let us know if your child is going to have any dental treatment. Sometimes, antibiotics are needed at the time of any dental work and also we need to check that their blood tests are satisfactory and that they are not on any drugs which might affect the dental work.



*Paulo and his brothers
beaching it*



Swimming pools and hot pools

Swimming is a great activity for children and it is important that all children learn to swim. However, we recommend that children do not go swimming in the first 3-6 months after liver transplantation. After this swimming in public pools is fine, as swimming pools should be chlorinated. The sea is fine (except for contaminated beaches!).

Hot pools are slightly different because the hot temperature means some bacteria (bugs) grow in them more easily. Older children who are more than 12 months post transplant can go in hot pools but must never put their head under water. We recommend that young children only go into hot pools if they are closely supervised by an adult and only then if they can understand that they must not splash around or put their head under water.



Hamish playing in the pool.

Holidays

Family holidays are important! We are very supportive of our patients and their families taking holidays. Short holidays

around New Zealand can be taken any time after 3 months post-transplant as long as we are able to contact you via cellphone while you are away.

We generally recommend that you do not travel overseas until after the first year. For some parts of the world, immunisations/vaccinations are required. Some of these vaccinations cannot be given to children on anti-rejection medications, which means that there are certain countries which your child will not be able to visit. Please ask us first before planning a holiday to a country where vaccinations are required or recommended.

Please ensure that you have good holiday insurance before travelling. If your child is unwell while you are away, it may be necessary to use medical facilities overseas or in serious cases be flown home to New Zealand. This can be extremely expensive unless your insurance covers it. We are happy to write a report for you to obtain insurance if necessary. The Kids Foundation has access to an insurer who will provide a quote based on a group rate through the foundation. You can contact IDFNZ for further details.

Please let us know of your travel plans early. We will then provide you with an open letter giving details of your child's history in case you need to visit a doctor while you are overseas. Please be aware that airline regulations will now only allow you to take liquids and liquid medications on board a plane if they are necessary during the flight. Liquids must be in small containers of less than 100 mls each and the total

amount of liquids must not be more than 300 mls. If you need to take liquid medications on the aeroplane, we will write a letter to cover this.

Pets

Children love animals and pets are allowed for transplant children, but some precautions must be taken. All pets must be vaccinated – this is very important. In addition, all animals should be wormed regularly. Young animals such as kittens hold the highest risk to transplant patients.

Reptiles and chickens often carry diseases such as Salmonella and do not make good pets for transplant recipients. If you are planning a new pet, especially a cat, you may wish to think about getting an animal that is slightly older and already vaccinated. It is important that children do not touch the litter trays of any cats and that they wash their hands after touching or handling the pet. The same rules apply if your child visits a petting farm.



Iharaia enjoying the swings at the park.

Child care arrangements

All parents worry about their transplanted child and many feel unable to leave them in the care of anyone else. However, parents need a break sometimes and we strongly recommend some time out for “you”. This may just be to do normal things such as getting a hair cut or shopping but please also remember to do things such as going out for a meal as a couple. Family members and friends are usually only too willing to help you by looking after your child while you go out. Make sure you leave a telephone number in order to be contacted in case of any queries or concerns.

Some parents choose not to allow carers to give medications and prefer to go out during times when medications are not needed. If you need to use a regular caregiver e.g. if you decide to go back to work, then we are more than happy to help that person learn medications, feeds etc. Please let us know if this is the case. We are also more than willing to write any letters of support that are needed if you require absence from work for appointments, admissions etc.

Stress

Transplantation is stressful for everyone involved. Sometimes parents “muddle through” the most frantic times when their child is most unwell and the time of transplant itself. It is often several weeks and months down the road that feelings of stress, anxiety and even depression become apparent (post-traumatic stress).

This is normal. Please let us help you if this happens to you. Many people just need someone to talk to, others may need more help or even medication. We are able to access the Consult Liaison service if this is necessary but some parents prefer to access their GP for further support and treatment. Please do not hesitate to let us know if you have any difficulties.

Influenza (‘flu) vaccination

We recommend that all transplanted children and their families receive a “flu vaccine every year. It usually becomes available in early autumn. The ‘flu virus is constantly changing and so the vaccine is different each year. This is why it is important to be vaccinated EVERY year.

Learning and implications

Researchers are increasingly concentrating their efforts on quality of life outcomes following liver transplant focusing on the impacts of transplantation, and potential strategies to manage these.



Ruby with hand picked flowers.



Utiku playing in the sand pit.

Other studies continue to work towards understanding the impact of liver disease in infancy on brain development. At birth almost all the neurons (brain cells) that the brain will ever have are present. However the brain continues to grow for a few years by the age of 2yrs the brain is about 80% of the eventual adult size. There are concerns that prolonged illness and periods of hospitalisation at this important stage of life may have serious life- long impacts.

Some initial findings have been presented, which suggest that approximately 1 in 4 children transplanted in infancy may develop learning difficulties ranging from mild to severe.

This is not an area which is completely understood as yet, however it is important to highlight so that families have every opportunity to get the right help from their school if their child is struggling for any reason.

It is important for us to know how your child is coping in school. If your child is having learning difficulties we can arrange for formal testing by a specialist service who can provide practical support and strategies for your child to manage their learning, and most importantly experience success at school.

Many people who have experienced learning difficulties in childhood have gone on to be incredibly successful adults with the right supports and strategies in place to help them.



Lara and Jordan at a Kids foundation family day – giving their parents heart attacks riding alone.



Family entitlements and Agencies to contact

There are a range of entitlements for families of sick children and families living away from home. These range from child disability allowance and carer support allowance to accommodation allowance and medical travel subsidy. Your social worker or Kids Foundation support person will point you in the right direction for all the support you can receive or you could contact Work and Income directly. Many of the forms will require the medical team to authorise.

Family networking

Many families find meeting another family/child in a similar situation both helpful and beneficial. You may meet other families on the ward when you are in-patient or your CNS or Kids Foundation support person can introduce you to other families on request. It is often helpful to meet a family 'further down the road' from you so you can see life will be normal again and that you/your child will be able to lead a normal life.

There is a Facebook support site (search for biliary atresia/liver transplant New Zealand) which has been set up by a mother of a transplant recipient. All pre- and post-



Emily, Samantha and Tracey with their children.

liver transplant families are welcome to join this online support group and network with other families around New Zealand to ask questions and gain valuable support and friendship.

The KIDS Foundation

The KIDS Foundation is the welfare arm of the Immune Deficiencies Foundation of New Zealand (IDFNZ). We offer support to children and adults with primary immune deficiencies, immune defects and some rare blood disorders. As part of KIDS we also offer support to children and their families who are on the liver transplant waiting list and who have undergone liver transplantation.

A group of parents started KIDS foundation in 1987. The foundation has the legal status of an incorporated Society and is managed by a National Executive Committee. The Foundation's work with Bone Marrow Transplant children led in 2002, to it taking on the responsibility to support immune compromised liver transplant children. IDFNZ is proud to be associated with the New Zealand Children's Transplant Support Trust (NZCTST) and Liver Transplant Children of New Zealand.

We know that, with the support of others who have experienced the ups and downs associated with the liver transplant process, you too can

make it through this challenging time. We hope you find both support and encouragement by being part of the KIDS Foundation.

Freephone 0508 300 600
www.idfnz.org.nz

Services offered

- Patient membership: Free membership, as it is our belief the price to be eligible to join is high enough.
- Support: we have a full time support person for families and patients.
- We can introduce families and organise get-togethers such as our annual Christmas Party and regular coffee groups.
- Education: we aim to provide informative brochures and information to families.
- We can offer some financial assistance to families, especially during the hospital stay in the form of car park vouchers and meal vouchers.
- Advocacy: with chronic illness it can feel like you are battling alone to gain quality of life. We have advocacy services available for you to use.
- We are committed to providing members with the latest information to empower families to make well informed life affecting decisions.

Nutrition after a liver transplant

In the months after a liver transplant it is important that your child moves towards eating a balanced healthy diet appropriate for their age. Those children who are eating relatively well before their transplant, generally eat well after transplant.

If your child has needed long-term tube feeding prior to the transplant, the transplant will not immediately solve their feeding difficulties. Over time your child will learn to feel hungry, to eat, drink and enjoy food after a transplant. A speech language therapist may help with this. Some children will need to go home with their nasogastric tube, particularly if they needed NG feeds before transplant.

Immediately after transplant

Your dietitian, after discussion with the transplant surgeons and gastroenterologists, will decide when feeding can commence after transplant. Tube fed children will continue on their pre-transplant formula in the short-term.

For the first few weeks after transplant, when your child's immunity is at its lowest as a result of their immunosuppressive medications, there is an increased risk of infection from food (food poisoning). You will be advised to follow the New Zealand Food Safety Authority handbook "Food Safety When You Have Low Immunity". Your dietitian will give you a copy at the time of your child's transplant (further copies are available from the information centre by the Outpatients Clinic on level 3 of Starship).

Following your child's transplant you can reduce the risk by:

- Good hand washing practice
- Ensure your child does not share knives, forks, spoons, bottles, cups and plates at meal and snack times.
- Your child should not share bites of food, licks of ice cream, packets of food etc with another person.
- If you would like to bring food into the hospital from home, please discuss this first with your dietitian.
- Ensure your child's food is eaten soon after it is cooked.
- Do not give your child café/deli/ takeaway food

Long-term after a liver transplant

Weight gain and catch-up growth occur after transplant. Your dietitian will review your child's growth and food intake at clinic.

Some medications, such as steroids, can result in excessive weight gain.

Children with normal liver function are encouraged to eat a variety of foods. Include foods from these 4 food groups to get the nutrients your child needs to grow and stay healthy:

- Milk and milk products
- Lean meats, chicken, seafood, eggs, peas, beans and lentils
- Bread and cereals
- Vegetables and fruit

CLEAN



WASH HANDS AND SURFACES OFTEN

SEPARATE



SEPARATE RAW MEATS FROM OTHER FOODS

COOK



COOK TO THE RIGHT TEMPERATURE

CHILL



REFRIGERATE FOOD PROPERLY

Complications after liver transplant

It is uncommon for a medical/surgical treatment as complex as a liver transplant in a child to be undertaken without some complication or other. Fortunately, the majority of complications which might be encountered are treatable.

These can be divided into groups as follows:

- Those specific to the surgery itself (mainly plumbing problems and graft problems)
- Those related to infection
- Those related to rejection
- Side effects of immunosuppression

Surgical plumbing problems and graft problems

Blocked Blood Vessels

Blocked blood vessels are diagnosed by a doppler ultrasound of the liver and are confirmed by angiogram. If detected early your child may need to undergo a re-operation to remove the clot. Usually IV heparin is used to thin your child's blood and prevent any further clot.

a) Hepatic Artery Thrombosis

Hepatic artery thrombosis (clotting of the main blood vessel that brings oxygenated blood from the heart to the liver) occurs in about 1-2% of all transplants (a bit higher in those who receive a living donor liver transplant).

The liver cells themselves typically do not suffer from losing blood flow from the hepatic artery flow (because they are also nourished by blood from the portal system), but the bile ducts depend strongly on the hepatic artery for oxygen and loss of that blood flow may lead to bile duct scarring and infection.

If this occurs it can sometimes be rectified with an operation to remove the thrombosis, but sometimes another transplant may be necessary.

b) Portal vein thrombosis

Portal vein thrombosis (clotting of the large vein that brings nutrient-rich blood from the intestines, the pancreas and the spleen) to the liver occurs rarely, mainly in certain cases of biliary atresia. This complication may or may not require a second liver transplant.

Bleeding

Bleeding is a risk with any surgical procedure, but a particular risk after liver transplantation because of the extensive nature of the surgery and because clotting depends on factors made by the liver. Most transplant patients have some bleeding during the surgery and may require transfusions before and sometimes after the operation. The bleeding may occur from a variety of sites, including the cut surface of a reduced size liver. If this happens, your child may have to return to the operating theatre to stop the bleeding. Usually bleeding will occur within the first twelve hours of the transplant. Your child will not be aware of this, as he/she will still be heavily sedated. About 5-10% of transplant recipients require a second operation to prevent recurrent bleeding.

Primary Non-Function

Sometimes the new liver does not work well from the get-go, a problem called primary non-function. This occurs in approximately 1% of new transplants. If the function of the liver does not improve sufficiently or quickly enough, the patient may urgently require a second donor liver.

Biliary complications

Biliary complications (leaks from or blockages of the bile ducts) are a bit more common, affecting approximately 15% of deceased donor whole graft transplants and up to 40% of partial liver donor transplants (split/cut down grafts).

Biliary leaks into the abdominal cavity occur either from where the donor bile ducts are connected to the recipient ducts or from the cut edge of the liver in living donor or split liver transplants. Typically, a drain is placed during the transplant operation along the cut edge of the liver to remove any bile that may leak. This drain will usually be removed within the first week after transplant. As long as the bile does not collect in the abdomen, the patient does not become ill. Leaks often heal with time, but may require additional treatment procedures.

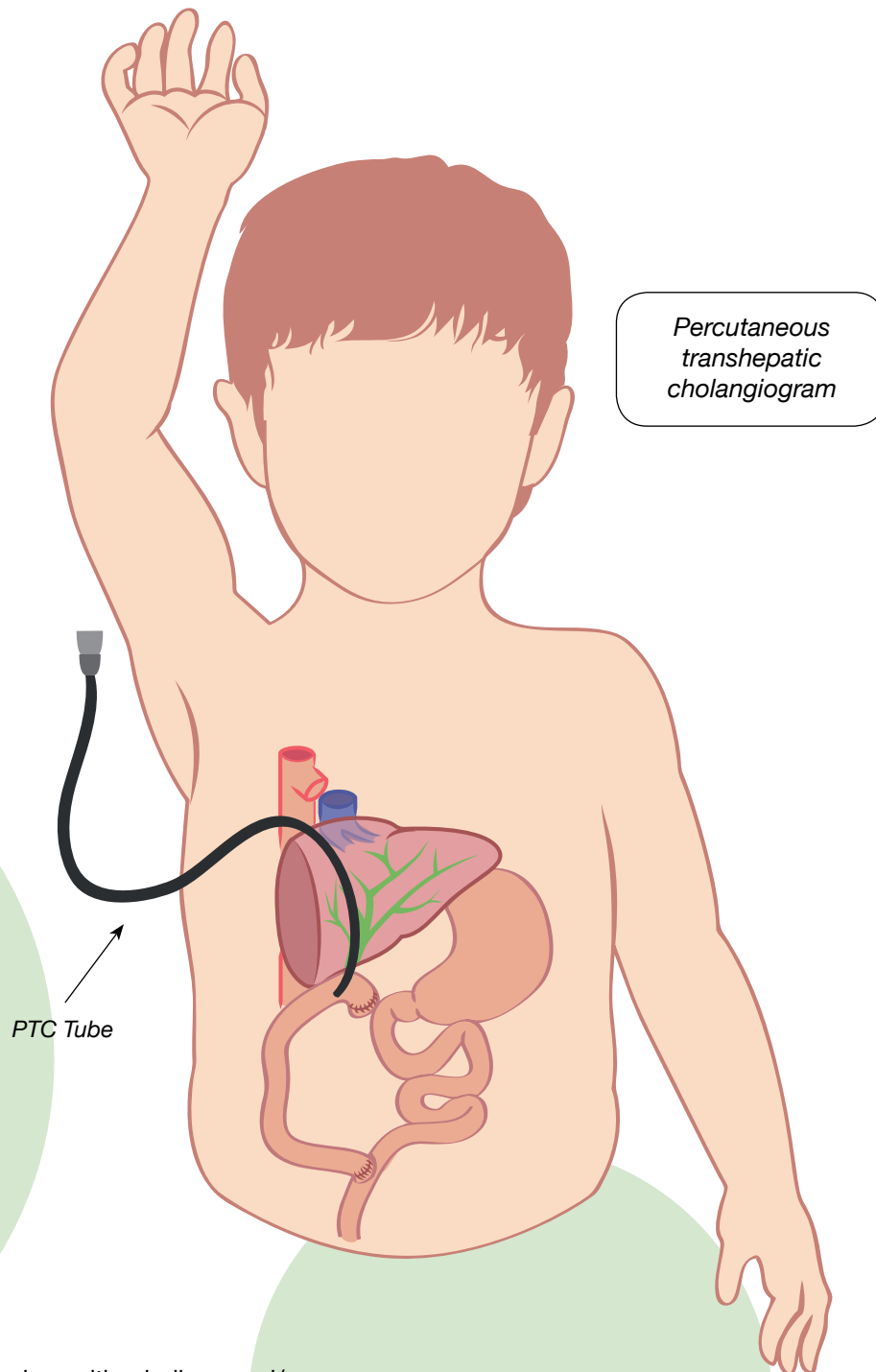
Biliary blockages (called strictures) result from the narrowing of the bile duct and relative or complete blockage of the bile flow. This may be accompanied by infection of the bile. Pain around the liver, abdomen and fever may be symptoms.

It is important to report any symptoms to the transplant team.

Strictures can appear weeks and sometimes months after the transplant. Most frequently, the narrowing occurs at a single site, usually where the donor and recipient ducts are sewn together. This is treated by a radiology procedure called a cholangiogram or PTC. A wire is inserted through the narrowing. The narrowed area is

stretched up with a balloon and/or a stent is placed across the stricture.

If these methods are unsuccessful, a further operation to connect the bile duct to a segment of intestine can be done. This is called biliary reconstruction surgery. Rarely, biliary strictures occur in multiple sites throughout the biliary tree. This may occur if the biliary tree was poorly preserved during the period when the liver was out of circulation or because of hepatic artery problems.



Infection

Infections are fairly common after liver transplants in children and can usually be treated with antibiotics, antifungals or antiviral medication. Your child's recent history of liver failure, as well as surgical complications such as bile leaks or abdominal collections, contribute to this risk initially.

As already outlined, the immunosuppressive drugs also weaken the transplant recipient's defences against infection, which means that your child will be more susceptible to infections, especially during the first three to six months. Transplant recipients can get the "normal" infections that may affect anyone, but also 'opportunistic' infections (infections which occur in people with compromised immune systems). The further out from transplant your child is, the less risk there is that your child will contract these infections.

To protect him/her from infection remember to use common sense:

- Avoid people who have infections (colds, flu, chickenpox, measles). If your child does come in contact with any of the above, inform the Transplant Team or your local physician.
- Ensure good personal hygiene, especially regular hand washing - using antibacterial soap for hand washing is a good idea throughout your home.
- Treat cuts and abrasions immediately by washing thoroughly with soap and water, applying an antiseptic cream and cover with a clean, dry dressing.
- Good oral hygiene with regular dental check-ups every six months after the transplant.

- Avoid swimming in public swimming pools for 3-6 months, hot pools for a year.

During the first month or two, infections with bacteria and fungi are most common, affecting up to 40% of children. In months 1-6, certain viral infections such as cytomegalovirus, and rarely, unusual infections such as tuberculosis and pneumocystis pneumonia may be seen. Later, as immunosuppression levels are reduced, exposure to everyday viruses occasionally lead to problems, the main ones being opportunistic responses to viruses such as chicken pox, cytomegalovirus (CMV) and Epstein-Barr virus (EBV) - the virus which causes glandular fever (infectious mononucleosis). We monitor carefully for exposure to these viruses, particularly EBV, which, if the infection takes hold, can develop into a nasty type of infection in the lymph glands called Post Transplant Lymphoproliferative Disorder (PTLD).

There are three main types of infection:

Bacterial Infections

Line Infections

The most common infections in children post-transplant are bacterial, often related to the central lines which are inserted at the time of the transplant. For this reason we retain these lines only as long as they are needed to deliver important drugs. These infections may be identified by culture of blood taken from these lines.

Wound infections

Wound infections cause swelling, redness, tenderness and ooze or drainage from the infected site. Other symptoms include lethargy and tiredness, fever and pain around the wound. Common sites of infection are the 'Mercedes' wound or wound drain sites. A swab will help to identify the exact bacteria causing the infection and appropriate antibiotics will be given.

“Around day five post-transplant Isabelle's dressing was removed to reveal she had a wound infection across her entire incision. Fortunately I wasn't there to see it, but her Dad was, and he nearly fainted.

Daily cleaning of the open, infected wound was needed for a while and holding her hands out of the way and comforting her while this was done was the one job I couldn't handle so I asked a nurse to step in. After a week or so the infection was looking much better and during a visit to theatre for something else the surgeon used external stitches to close the wound.”

– Jodie, mother of Isabelle

Chest infections

Chest infections can be due to many different bacterial and fungal infections. Symptoms may include a dry or productive cough (coughing up phlegm), fever and rapid rate of breathing. A chest X-ray may help to confirm the diagnosis. Antibiotics or antifungal medicines will be used to treat the infection.

Viral Infections

Cytomegalovirus - CMV

This is the most common viral infection your child is likely to encounter. The risk of CMV is highest within the first six months post transplant. Symptoms include general fatigue, diarrhoea, raised temperature, flu-like symptoms and pneumonia. If your child has not had prior exposure to the infection he/she will be prescribed a drug called Valganciclovir to take for the first 3 months post-transplant. This is usually given either as a tablet form or a liquid preparation made up in the hospital pharmacy.

Epstein Barr Virus EBV

EBV infection can be a significant long term problem in children after transplant. The majority of young children have not had this infection prior to transplant, so will have no immunity to the virus and will be susceptible to infection. We regularly test for this infection (via blood tests) and aim to reduce immunosuppression if viral levels become significantly elevated. This is to prevent post-transplant lymphoproliferative disease (PTLD). We also use Valganciclovir to prevent infection with this virus early after transplant.

Post-Transplant Lymphoproliferative Disorder (PTLD)

PTLD can occur in approximately 1-2% of post liver transplant patients. It is almost always associated with EBV infection, the same virus that causes glandular fever or 'the kissing disease'. EBV infection is very common and the majority of adults (approximately 95%) have been exposed to EBV, most commonly in their childhood or teenage years. Most children transplanted before the age of 2 years come to liver transplantation without ever having been exposed to EBV. As it is so common, children will inevitably be exposed to EBV after transplantation. Under the influence of high levels of immunosuppression, they may be unable to control the infection, in this situation the infection can evolve into a form of pre-malignant lymphoma (cancer of the lymph glands).

As it is a result of a compromised immune system, the first line of treatment is to simply reduce and in some cases stop the immunosuppression. This allows the body's immune system to fight off the infection. This frequently works without incident, but there is a risk of graft rejection which might then require an increase or change in immunosuppression treatment.

Under these circumstances a common approach is to reduce levels of immunosuppression, but also give a medicine called Rituximab. Rituximab specifically eliminates B cells (the type of white blood cells infected by EBV). If this approach does not work, then more conventional lymphoma

cancer chemotherapy may be used. Although it is a serious and worrying complication of transplant, the majority of PTLD cases can be successfully treated.

Chicken Pox (Varicella Zoster)

The issue of chicken pox is one that can cause families significant anxiety, due to its high incidence in the community. The following section attempts to take away the mystery of what constitutes an exposure, why we take it so seriously, and what the likely treatment will be.

Chicken pox is a common childhood infection, which generally causes an unpleasant, but relatively short-lived illness in most children. In children who are immunosuppressed, a more severe 'life threatening' infection may occur. If you suspect your child has been in 'direct' contact with chicken pox, we strongly advise you to contact the transplant team for advice and a management plan.

In immunocompromised children, progressive severe chicken pox (varicella) may be characterised by:

- continuing eruption of lesions
- high fever persisting into the second week of illness
- encephalitis (swelling of the brain)
- hepatitis (inflammation of the liver)
- pneumonia (chest infection)

Infection occurs when the virus comes in contact with the mucosa of the upper respiratory system or the conjunctiva (surface of the eye). Person to person infection occurs mainly by direct contact (ie being in the same room/breathing the same air for more than 5 minutes) with a person with active infection.

Chicken pox symptoms

The most commonly recognised chicken pox symptom is a red rash that can cover the entire body.

Before the rash appears, your child may have mild flu like symptoms which include:

- feeling sick
- a high temperature (over 38 C)
- aching, painful muscles
- headache
- generally feeling unwell
- loss of appetite.

Soon after the flu-like symptoms, an itchy rash appears, some children may only have a few spots, but others are covered from head to toe. The rash starts as small itchy spots, after 12-14hrs the spots develop a blister on top and become incredibly itchy.

The spots normally appear in clusters and tend to be

- behind the ears
- on the face
- over the scalp
- under the arms
- on the chest and belly
- on the arms and legs

Spots can be anywhere on the body, even inside the ears and mouth, on the palms of the hands, soles of the feet and inside the nappy area.

Defining 'Chicken pox'

Contagiousness:

Chicken pox is most contagious 1 to 2 days before to shortly after the onset of the rash. Contagiousness persists until crusting of the lesions occurs.

Incubation Period: Usually 14 to 16 days after contact, can be as long as 28 days after the use of VZIG (Varicella-Zoster Immune Globulin)

“ One of my biggest nightmares has been the constant rounds of chicken pox in our community. We were constantly vigilant though and after being told by a friend of Lara’s exposure to chicken pox, when we saw a suspicious spot appear we called our team at Starship and let them know we were on our way in. They were well prepared for us and by the time we made it to hospital a couple more spots had appeared confirming our diagnosis. Lara was admitted immediately and was started on an anti-viral treatment. Because of our quick response we only stayed a couple of miserable days and Lara had only a few spots and no scars. She still has immunity to chicken pox at this stage too. We have also had two other previous trips to hospital with suspected exposure and stayed on a couple of hours to receive immunoglobulin as a precaution. Keep vigilant and you will be ok! ”

– Samantha
mother of Lara

Action Plan for contact with Chicken Pox:

At initial discharge post-transplant, we will advise you as to whether your child has immunity to chicken pox (due to previous infection or immunisation). This level of immunity will be checked yearly on blood testing, as immunosuppressed children can lose this protection over time.

If your child has no protection against Chicken Pox:

Contact CNS, with a description of the exposure.

We will want to know:

- Is the exposure from a sibling in the same house?
- Was the contact with a playmate (face to face indoor play for longer than 5 minutes)?
- Has there been possible Hospital Inpatient exposure?

If it is within 96hrs of known exposure –we will request that your child receive a dose of VZIG at your local hospital. VZIG gives your child a degree of temporary protection against the chicken pox virus.

If outside the 96hr period your child will be prescribed oral Acyclovir 4-5 times daily for 7 – 10 days.

It is still possible for your child to develop chicken pox, so we would ask you to be vigilant in checking your child for possible signs of symptoms of chicken pox over the known incubation period.

If your child develops chicken pox, they will need to be hospitalised to receive IV Acyclovir (anti viral medicine) to reduce the possible effects of the illness. Isolation precautions will be maintained for the period of the hospital stay.

So: your child can have chicken pox. However, it is vital they receive treatment early during the illness to prevent the ‘severe form of chicken pox’.

Vaccination cannot be given soon after transplant, as the chicken pox vaccine is a “live” vaccine. However, it may be considered when your child is considered a safe period of time post-transplant. We will still monitor your child’s immunity to chicken pox, as they may lose this immunity over time.

Mumps

Prior to the introduction of the MMR vaccination in 1969, Mumps was a very common childhood infection. It is now rarely reported and is mainly seen in children aged between 5 and 14 yrs. It is caused by a virus that usually spreads through saliva (spit) and can infect many parts of the body, especially the parotid glands located between the ear and jaw, normally the virus causes these glands to become swollen and painful. In children who are immunosuppressed, we can anticipate a more severe form of the infection occurring if they are exposed to the virus. If you suspect your child has been in 'direct' contact with mumps, we strongly advise you to contact your paediatrician or the Transplant team for advice and a management plan.

Measles

Measles is a notifiable highly infectious illness that can cause fever, coughing and distinctive red-brown spots on the skin. Measles is preventable through childhood vaccination. Every effort is being made in New Zealand to boost rates of immunisation. Until we reach 95% coverage we will continue to see outbreaks of measles in the community. Unfortunately many children who have required transplantation in infancy were too unwell to receive the vaccination pre transplant, therefore they are vulnerable when there are measles outbreaks in the community.

Measles is spread by droplet, with the virus remaining viable (alive) for 2 hrs outside the body. Any patient with measles will require immediate

isolation, both in the hospital and community. The public health team will request that people with active cases of Measles remain in their home.

Measles in severe form (1:1000 cases) can cause swelling of the brain (encephalitis) which can lead in extreme cases to a persistent vegetative state.

The initial symptoms of measles include:

- Cold-like symptoms such as a runny nose, watery eyes, swollen eyelids and sneezing
- Red eyes and sensitivity to light
- A mild severe temperature, which may peak at over 40.6 C for several days, then fall but go up again when the rash appears
- Tiny greyish-white spots (called Koplik's spots) in the mouth and throat
- Tiredness, irritability and general lack of energy
- Aches and pains
- Poor appetite
- Dry cough
- Red- brown spotty rash.

Defining 'Measles' Contagiousness:

Measles is most contagious 1 to 2 days before the onset of the symptoms persisting for approximately 5 – 9 days.

Incubation Period: Usually 7 to 18 days after contact, can be as long in a child who is immunosuppressed.

Action Plan for contact with Measles:

At initial discharge post-transplant, we will advise you as to whether your child has immunity to measles (due to immunisation or in some cases exposure to the virus). This level of

immunity will be checked yearly on blood testing, as immunosuppressed children can lose this protection over time.

If your child has no protection against measles:

Contact CNS with a description of the exposure.

We will arrange for immediate assessment and for your child to receive an immunoglobulin infusion if the contact has occurred within 6 days of notification.

For unvaccinated family members we recommend that they are immunised within 3 days of contact as this will reduce the rate of secondary cases by approximately 80%

If your child develops measles they will require hospitalisation for treatment.

Any susceptible contacts will require Isolation/exclusion for 14 days from the date of the last exposure to an active case of measles (this is a public health measure).

Vaccination for Measles, Mumps, Rubella and Chicken Pox post transplant:

Vaccinations that protect against these illness are live vaccines meaning that in an immunosuppressed patient they can sometimes cause the illness to occur.

In a child who is highly immunosuppressed we would not advise that they have either the MMR vaccine or the Varicella vaccine for Chicken Pox. However in certain circumstances it may be felt it advantageous for your child to receive these vaccines. The pros and cons of this will be discussed with you fully with the final decision being a family decision.

We attempt to update families yearly of their child's immune status for the following

Hepatitis A and B; Chicken Pox and Measles, and will provide you with appropriate advice according to their time post transplant.

Fungal Infections

Candida (Thrush)

This can be a severe infection in transplanted patients. It can occur in the mouth and throat as white patchy areas. We use the medication Nystatin to prevent this infection and in some high risk situations we will use Liposomal amphotericin to prevent this.

Pneumocystis Jiroveci Pneumonia (PCP)

This is a germ similar to a fungus. In people whose immune systems have been suppressed it can cause a type of pneumonia (PCP). To prevent this

infection your child will receive Co-trimoxazole tablets or liquid taken three days per week for twelve months after transplant. Early symptoms of this infection include a mild, dry cough and, possibly fever.

If you suspect any type of infection in your child within the first three months, you must contact your transplant team as soon as possible.

After this period, your child's local paediatrician can be contacted.

Rejection

Why does the body reject the new liver?

What are the signs and symptoms of rejection?

Rejection is the term we use when there is inflammation of the donor organ caused by the recipient's immune system reacting to the transplanted organ. The immune system consists of specialised white blood cells which fight infectious organisms, such as bacteria and viruses. When your child comes in contact with an infection, these blood cells rush to the site and surround the invading organism. The body then produces antibodies which destroy the organism and render it harmless.

When a new liver is transplanted, these white cells view it as an invading organism and may then attack it. This is a normal reaction. To reduce the risk of this happening, the immune system must be suppressed. This is done by giving immunosuppressive (or anti-rejection) drugs. However, because these alter the immune system's response to invading organisms your child will be more susceptible to infections. It is important to get the right balance in giving enough medication to prevent the body rejecting the new liver, but not too much which will increase the risk of infection.

1) Acute rejection

When transplanted the body always recognises the new liver as "foreign" and tries to reject it to varying degrees. The risk varies depending on the age of the patient (children under 2 years have a lower risk) and the degree to which we use and monitor immunosuppression.

Episodes of rejection occurring within the first weeks or months post-transplant are generally termed acute rejection. Fortunately, in liver transplantation, unlike other organs, acute rejection does not generally affect overall chances for graft survival. This is believed to be because the liver has the unique ability to regenerate when injured there by restoring full liver function.

Although episodes of acute rejection are fairly common (approximately 50% of children have at least one episode), the highest risk is only in the first 4-6 weeks. After this time the risks of rejection become less, but rejection can occur at ANY TIME, even many years later. If you reduce or stop your medication rejection will occur.

Usually acute rejection occurs without any signs or symptoms. Patients do not feel any different or notice anything. The first sign is usually abnormally elevated liver tests.

Occasionally rejection may cause:

- Irritability
- Fatigue or general tiredness
- Poor appetite
- Flu-like symptoms
- Jaundice or yellow colouring of the eyes and skin
- Fever
- Itching
- Light-coloured stools
- Dark-coloured urine
- Tenderness or pain in the abdomen
- Abdominal swelling

Rejection is always diagnosed by a liver biopsy. This is usually performed by a radiologist using ultrasound guidance while your child is anaesthetised.

It is important to contact the transplant team if you note any of the above symptoms in your child after he/she has been discharged home.

Do not wait until the next outpatient appointment.

Once the diagnosis is made, treatment is fairly straight forward and generally very effective. The first line of treatment is high dose corticosteroids (see 'medications'). Over 90% of rejection episodes can be treated effectively with an increase in medication which may include:

- An increase in your daily immunosuppression drug.
- A three-day course of intravenous steroids (Methylprednisolone).
- More potent immunosuppression drugs, eg ATGAM or OKT3

A small proportion of acute rejection episodes (approximately 10%) do not respond to corticosteroid treatment and require additional treatments.

2) Chronic Rejection

Chronic rejection occurs in less than 2-3% of liver transplants in children, particularly in those with repeated episodes of acute rejection. Liver biopsy shows loss of bile ducts and obliteration of small arteries. In the past, chronic rejection proved very difficult to reverse, often causing graft failure. However, today, with our large selection of immunosuppressive drugs, chronic rejection is more often reversible.

Diabetes

Some of the immunosuppressive medications, such as Prednisone or Tacrolimus, can cause an increase in blood sugar levels resulting in diabetes. This is usually temporary and resolves when your child's Prednisone dose is reduced. It may occasionally be necessary to test his/her blood sugars levels by regular finger pricks and have insulin injections. If this should occur, you will receive specialised teaching about how to deal with this problem.

Hypertension

Hypertension (high blood pressure) may occur as a side effect of immunosuppressive medications such as Tacrolimus, Sirolimus or Prednisone. Your child's blood pressure will be monitored closely on the ward and then at each clinic visit. If it is necessary, he/she may be commenced on a drug to lower his/her blood pressure. This is often a problem in the early weeks post transplant, but resolves when the doses of prednisone and/or Tacrolimus are reduced.

It is possible that as part of your child's annual review that they may undergo 24hr Blood pressure (BP) monitoring. They will be able to move around as normal but will have the inconvenience of wearing a monitor which takes BP measurements at regular intervals over the 24 hrs.

Medications

What are immunosuppression medications?

Immunosuppression medications are commonly used drugs which help prevent rejection by suppressing the immune system. They work through different mechanisms to weaken the immune system's response to stimuli and are associated with different side effects.

As a result, these medications are frequently used in various combinations which increase the overall immunosuppressive effect while minimising side effects.

Current practise is to use a combination of 2-3 of these medications in varying doses for the first year after a liver transplant. For example, your child may receive Prednisone + Tacrolimus, or Prednisone + Tacrolimus + MMF. However, most patients will be weaned to Tacrolimus alone after the first year post-transplant. These variations will be tailor-made to each patient according to their complications and needs.

As previously stated, the immune system's primary role is to identify and attack anything that is foreign or non-self. The main targets were not intended to be transplanted organs, but rather bacteria, viruses, fungi and other micro-organisms that cause infection.



The most serious side effect of immunosuppression is infection as mentioned above. However, in addition to fighting infection, the immune system has a role in fighting cancer. It is believed that a healthy immune system detects and eliminates abnormal, cancerous cells before they multiply and grow into a tumour.

It is well recognised that transplant recipients are at increased risk for developing several specific types of cancers, particularly skin cancers, with a 25-fold increase in risk compared with the normal population. In view of this substantial risk, it is strongly recommended that all transplant recipients minimise sun exposure.

What medicine will your child be on?

Standard Medicines at discharge post transplant are listed below:

Initially your child will be on a wide range of medicines ranging from immunosuppressive medicines

(antirejection), steroids, antifungals, antivirals, antibiotics and medicines that lower your child's blood pressure.

They all play a very important part in preventing both rejection and infection, and combatting side effects of other medicines. It is important to report any side effects to the transplant team.

The most commonly used medicines are each identified with an *. It is important to remember that everyone is treated individually and therefore may require a different combination of drug therapy.

Steroids

Corticosteroids are a class of anti-inflammatory agents that inhibit production of cytokines, the molecules produced by cells of the immune system which orchestrate and intensify the immune response. They prevent activation of lymphocytes, the main soldiers of the immune response against transplanted organs. There are 2 main forms of steroid medicine: methylprednisolone is given intravenously and prednisone is given orally.

Methylprednisolone (methylPRED) is given intravenously during and just after a transplant, until oral medications can be administered. In addition, if there are any later episodes of rejection, this intravenous form of steroid hormones may be given for three days in high doses.

Prednisone *

Prednisone (PRED) is used in regular low doses as a preventative in the first 6-12 months following transplant. High doses will be used immediately after the transplant, but the dose

will gradually be reduced to a lower maintenance dose and may be withdrawn at one year after transplant. If your child's original liver disease was Autoimmune Hepatitis, they will remain on a low dose.

- Prednisone is taken once a day, preferably in the morning.
- It comes in 1mg, 2.5mg, 5mg, and 20mg tablets.
- It is also available as an oral solution as prednisolone 5mg/ml (Redipred® -cherry flavour). This is essentially the same as prednisone.
- Prednisone should not be taken on an empty stomach, always give to your child with a feed or food.
- Prednisone should NEVER be stopped suddenly.

Side effects include fluid retention, raised blood pressure, an increase in appetite causing weight gain, stomach ulcers, skin changes (such as papery skin, acne, bruising, delayed healing), increased body hair, sleep disturbance, mood swings and diabetes. Most of these side effects resolve when your child's dose of Prednisone is reduced or can be combatted with other medicines.

Tacrolimus *

Tacrolimus is called TAC or Prograf® and is the most common drug prescribed to prevent rejection. Your child will almost certainly be on this drug for life. Your child will need to have regular blood tests to measure the level of the drug in their blood and to monitor and detect any side effects. The amount your child will need to take will alter depending on their blood results.

Tacrolimus belongs to a group of drugs called Calcineurin inhibitors – These drugs, were first developed approximately 20 years ago and revolutionised organ transplantation. They substantially reduced the incidence of rejection, improved the longevity of transplanted organs and thereby improved the overall success of transplantation. Unfortunately these drugs do come with significant potential side effects.

- Tacrolimus is taken twice a day, 12 hours apart.
- It comes in 0.5mg (yellow), 1mg (white) and 5mg (pink) capsules. **It is very important not to mix these up.**
- If your child is too young to swallow capsules, you will be shown how to mix the contents of the capsules in water before it is given. (Please see the next page for guidelines).
- Your child will need to take Tacrolimus on an empty stomach and be nil by mouth 1 hour before the dose and 1 hour after the dose.

The dose must be given at the same times each day.

- Your child should not eat grapefruit, or drink grapefruit juice, as this may increase the blood levels of the Tacrolimus. Always check with your doctor or pharmacist before giving your child any other medicines (including ones you might buy from your local pharmacy).
- Capsules should be stored at room temperature and in a safe place. Keep the capsules in the blister packaging until it is time to use them.
- **On the day of your child's blood test, do not give them their morning dose of Tacrolimus until after their blood test.**
- You must never alter your child's drug dose. The transplant team or your local paediatrician will advise you on this as necessary.

Side effects can include headaches, high blood pressure, tremor, insomnia (poor sleep pattern), abnormal kidney function, hair loss, diarrhoea, diabetes and sometimes seizures. Usually these side effects are related to higher doses and can be minimised by reducing the dose. The side effects lessen after the first month. Another side effect is light sensitivity. Please protect your child when in the sun, apply sunblock.

Instructions for making up a Tacrolimus dose from capsules

This is a step by step guide for parents/caregivers to make up a Tacrolimus dose for children who are too young, or are unable to swallow the capsules whole.

To make up the Tacrolimus dose you will need.

- Clean Hands
- Tacrolimus capsules
- Clean gloves (optional)
- Suitable dedicated container
- 3ml syringe
- Fresh tap water

1. Wash and dry hands thoroughly
Put on gloves.
2. Draw up 1ml of water into 3ml syringe.
3. Put 1ml of water into the container.
4. Hold Tacrolimus capsule upright in one hand and gently tap to allow powder inside capsule to settle in the bottom part of the capsule.
5. Gently pull top part of the capsule off and tip the powder into the container and gently mix using the syringe. **If any Tacrolimus powder spills discard the capsule and start again with a new one.**
6. Draw up all the Tacrolimus into the 3ml syringe and gently turn the syringe to ensure the Tacrolimus and water mix together.
7. Hold the syringe upright and gently push the plunger upward to dispel all the air and the top of the black plunger is at the 1ml mark on the syringe.
8. To measure the dose:
1ml = 1mg of Tacrolimus
0.1ml = 0.1mg of Tacrolimus

Examples

- If your child's dose of Tacrolimus is 0.3mg you will need 0.3ml.
- Gently push the plunger until the top of the black plunger is at the 0.3 mark on the syringe. Discard the rest of the medication you do not need down the sink and flush with water.
- For doses greater than 1mg, mix with a larger volume of water, e.g. 2mg in 2ml, 3mg in 3ml. If the dose is a fraction of 1mg, e.g. 1.6mg, use 2 X 1mg capsules in 2ml (= 1mg per 1ml) and give a 1.6ml dose.
- Note that 0.5mg capsules will require 0.5mls of water.

It is best to prepare this medicine well away from food preparation areas, just to be safe.

Remember that there is also a 5mg-sized capsule available in NZ. It is a pink colour. Always make sure you are aware of the dose and capsule strength you are working with:

0.5mg = yellow

1mg = white

5mg = pink

Mycophenolate Mofetil (MMF, Celcept®)

Mycophenolate is another medicine that is used to prevent or treat rejection which has not responded to high-dose steroids.

It can have a lot of side effects.

Mycophenolate mofetil dampens the immune response by preventing proliferation of lymphocytes. The primary side effects of mycophenolate mofetil affect the intestinal system resulting in stomach upset and/or diarrhoea. It can also depress bone marrow function and thereby, reduce blood levels of white cells (infection fighting cells), red cells (oxygen carrying cells) and platelets (clotting agents).

- Mycophenolate is usually taken twice daily.
- It comes in 250mg capsules and 500mg tablets. It is also available in an oral suspension 1g/5ml.
- It is given as well as Tacrolimus.
- Do not open the capsules, swallow whole.
- Mycophenolate can be taken with food to reduce gastric side effects.
- Sometimes the dose is split into smaller doses to reduce gastric side effects.

Side effects include nausea, vomiting, diarrhoea, constipation and anorexia. Other side effects are anaemia (low red blood cell count) and low white blood cell count. Regular full blood counts will be carried out to monitor for such problems.

Cyclosporin (Neoral®)

Some patients may be changed to cyclosporin as their antirejection medicine. This is also a calcineurin

inhibitor, like Tacrolimus, but it has a slightly different side effect profile.

- Cyclosporin is taken twice a day, 12 hours apart.
- It comes in 25mg, 50mg, and 100mg capsules and as an oral solution 100mg/ml.
- The oral solution should be used within 2 months of opening the bottle and stored between 15 and 30°C
- Cyclosporin may be taken with or without food.
- Your child should not eat grapefruit, or drink grapefruit juice, as this may increase the blood levels of the Cyclosporin.
- On the day of your child's blood test do not give them their morning dose of Cyclosporin until after their blood test if the trough level is being measured. Sometimes, levels 2 hours after a dose are measured (known as C2 levels) so doses will need to be taken 2 hours before the blood test.
- The oral solution can be diluted with orange or apple juice in a glass container immediately before taking to disguise the taste. Always dilute the dose in the same way.
- Refer to the product information sheet for instructions on how to measure Cyclosporin oral solution.

Side effects include irritability, increased blood pressure and hair growth. It may also cause gum enlargement and abnormal kidney function. Usually these side effects are related to higher doses and can be minimised by reducing the dose. Another side effect is light sensitivity. Please protect your child when in the sun, apply sunblock & avoid sunbathing.

Sirolimus (Rapamune®)

Very occasionally patients may be changed to Sirolimus, or it may be added to Tacrolimus. This is an anti-rejection drug.

- Sirolimus is usually taken once or twice a day.
- It comes in 1mg, 2mg tablets and as an oral solution (1mg/ml)
- The solution must be stored in the fridge
- Doses must be given consistently at the same time in relation to food each day.
- Your child should not eat grapefruit, or drink grapefruit juice, as this may increase the blood levels of the Sirolimus.
- On the day of your child's blood test do not give them their morning dose of Sirolimus until after their blood test.
- The oral solution should be diluted with water or orange juice immediately before taking – do not mix with any other liquids

Side effects include poor wound healing, low white blood cell count and an increase in blood lipids and cholesterol. Regular blood tests are necessary to monitor blood levels of the drug and any side effects. Another side effect is light sensitivity. Please protect your child when in the sun, apply sunblock & avoid sunbathing.

Please note if your child develops mouth ulcers it may be necessary to change to twice daily dosing.

Important Medicine Reactions

Are there any medicines which might cause problems after transplant?

You should not give your child any medicines which are not prescribed by your doctor. Some herbal or over the counter medicines can be dangerous to your child's transplanted liver or can have adverse reactions to the immunosuppression medicines which they are taking.

- Before using **ANY** other medicines for your child, please ask your transplant team first.
- Never stop or change the dose of your child's medicine without your doctor's approval.
- If your child requires any painkillers, use Paracetamol at recommended doses appropriate for their age and weight. As with any pain, if it persists or worsens, take your child to see your doctor. Avoid Aspirin and nonsteroidal anti-inflammatory medicines (NSAIDS) such as Ibuprofen (Nurofen, Brufen), Diclofenac (Voltaren), or Naproxen (Naprosyn) unless prescribed by the transplant team – they increase the likelihood of side effects of immunosuppression.
- Discuss any alternative/ complementary therapies with the transplant team before using them, as they may increase the likelihood of side effects of immunosuppression.

IMPORTANT REACTIONS to TAC, SL, or Cyc:

Many medicines or herbal remedies can interact with Tacrolimus, Sirolimus or Cyclosporin. Please inform the Liver Transplant team if your child is prescribed any of the following listed medicines:

The medicines listed below may **REDUCE** blood levels of immunosuppressants.

- Rifampicin
- Carbamazepine
- Phenobarbitone
- Phenytoin
- Prednisolone
- St. John's Wort

The medicines listed below may **INCREASE** blood levels of immunosuppressants.

- Grapefruit juice
- Clarithromycin
- Itraconazole
- Roxithromycin
- Fluconazole

Other Drugs

- Ketoconazole
- Voriconazole
- Metoclopramide
- Erythromycin

- Metronidazole
- Omeprazole
- Oral contraceptives

Ranitidine

Ranitidine combats stomach irritation and ulcers that may be a side effect of Prednisone or Aspirin. It is started at the time of transplant intravenously and then converted to an oral dose once your child is able to take medicines orally. It is usually continued for 3 months until the prednisone is reduced to a lower dose.

- Ranitidine is taken twice a day, in the morning and in the evening.
- It comes as 150mg tablets and as an oral solution 15mg/ml. The tablets can be cut in half.
- Ranitidine can be taken with or without food.

Co-Trimoxazole* (Sulphamethoxazole/ Trimethoprim)

Co-trimoxazole is given to prevent PCP (Pneumocystis Jiroveci), a type of chest infection which may occur in some immunosuppressed children. It is commenced after transplant and is continued until one year post-transplant.

- Co-trimoxazole is taken twice a day, three times a week on Mondays, Wednesdays and Fridays only.
- It comes in 480mg tablets and as a oral suspension 240mg/5ml.
- It should be taken with food.

Nystatin (Nilstat®)*

Nystatin is a oral liquid given to prevent fungal infections in the mouth such as thrush. It is used for three months after transplant.

- Nystatin liquid is given four times a day.
- It is a cherry flavoured yellow

suspension.

- The liquid should be swished around the mouth for around a minute, before swallowing. If your child is very young, you can use your finger to spread the liquid around their mouth.
- Your child should not have anything to eat or drink for at least half an hour after giving nystatin.

Folic Acid

Folic acid is used as a supplement to help produce healthy blood cells in the first three months after transplant.

- It comes as 5mg tablets.
- These tablets can be cut in half, crushed and mixed with water to give to your child.

Valganciclovir (Valcyte®) *

Valganciclovir is the medicine given to treat or prevent CMV and EBV infection (cytomegalovirus/Epstein Barr Virus). It is usually used for three months after transplant to reduce to risk of infection. It may also be used at other times to treat active infection.

- It can be given intravenously (ganciclovir) or as a tablet (valganciclovir).
- It comes as 450mg tablets.
- Tablets should be taken with food.
- If your child is too young to swallow tablets or requires a small dose, you will be shown how to crush and mix the tablet in water before it is given. (Please see the next page for administration guidelines).

As Valganciclovir can cause birth defects, all patients, male and female, should use adequate contraception whilst they are taking this drug and for three months thereafter.

Side effects include reduced white blood cell count (which may make your child more susceptible to infections),

abnormal kidney function, headaches, fever, rash and irritability. In some cases it can reduce your child's platelet count, which may make them more prone to bleeding. You should inform the Transplant Team if you notice any unusual bleeding or bruising, black tarry stools or blood in your child's urine.

Where possible you will be provided with the liquid formulation of Valganciclovir.

Instructions For Making Up A Valganciclovir Dose From Tablets

This is a step by step guide for parents/caregivers to make up a Valganciclovir dose for children who require small doses too young, or are unable to swallow the tablets whole.

To make up the Valganciclovir dose you will need:

- Clean Hands
- Valganciclovir tablet 450mg
- Clean gloves (optional)
- Tablet cutter
- Tablet crusher/pulveriser
- 3ml syringe
- Fresh tap water

1. Wash and dry hands thoroughly.
2. Put on gloves (and mask if preferred)
3. Draw up 1 to 2ml of water into 3ml syringe.
4. Using a tablet cutter, cut valganciclovir tablet in half (225mg) and in quarters (112.5mg) to give desired amount for your child's dose
 - 337.5mg is 3 quarters of a tablet (one half and one quarter)
 - 225mg is half a tablet
 - 112.5mg is quarter of a tablet
5. Place the required half or quarter into the bottom part of the tablet crusher. Place the lid of the tablet crusher on top and turn the device to crush the tablet. Continue to do so until the contents of the tablet are well crushed.
6. Carefully remove the lid of the tablet crusher, being careful not to spill any of then contents.

7. Put water into the tablet crusher to mix the powered tablet. Gently mix the crushed tablet with water using the syringe and remember to rinse any powder off the lid of the tablet crusher into the mixture.
8. Draw up all the valganciclovir into the 3ml syringe and gently turn the syringe to ensure the valganciclovir and water mix together.
9. Ensure all the valganciclovir is drawn out of the pill crusher. If powder remains in the crusher, used liquid in syringe to rinse pill crusher and draw up mixture again. This may take several attempts.
10. Hold the syringe upright and gently push the plunger upward to dispel all the air.
11. Give the dose to your child preferably with food.
12. Once you have given the dose, carefully rinse the syringes and contained, wipe down the work area, and wash your hands. It is best to prepare this medicine well away from food preparation areas, just to be safe.

Aspirin

Aspirin is used in low doses to thin the blood post-transplant to prevent any clots in the blood supply to the new liver. It is usually continued for three months.

- It comes as a 300mg soluble tablet that can be dispersed in water.
- Other forms of aspirin include 75mg and 100mg enteric coated tablets.
- Doses should be taken with food.
- If your child is charted a small dose, disperse one 300mg tablet in 10ml of water and mix well. This gives a 30mg/ml solution. For example to give a dose of 37.5mg you will need to give 1.25ml of this solution.
- Discard remaining solution you have made after each dose.

Side effects include increased risk of bleeding and stomach irritation. You should inform the Transplant Team if you notice any unusual bleeding or bruising, black tarry stools or blood in your child's urine.

Anti-Hypertensives

There are many anti-hypertensive medicines, which can be used to lower your child's blood pressure. Your child may already be on one or more of these prior to transplantation, or may be commenced on these due to side effects of the immunosuppressive medicines.

Life accounts



“ Paulo was diagnosed with Biliary Atresia at the age of 6 weeks. From that day, we were in what felt like a long physically, mentally and traumatic roller coaster ride.

With the help, love and assurance of our friends and family and the utmost professionalism, dedication and caring support of the Liver Transplant team this country has to offer our ride was a lot more bearable!

We are thankful every day to have Paulo, who is now 7 years old, fit, healthy and full of life and an absolute character. ”

– Phil, Frances, Rio, Gabriel and Paulo



“ When I look at Alice today I can't believe that she went through this experience. She is a very normal, bubbly, tantrum-pulling 2 year old. I wish I'd had this image 2 years ago when at aged 4 months she was given a new liver. The medical team at Starship saved her life; we are forever in their debt.

Some nights were very long and lonely; you'll be very surprised how strong you can be when you have a sick child. And it doesn't last forever, there is an end point. Lean on family and friends, support will get you through. No-one can do this journey alone.

If you can, talk to other families who have been through this. It's so reassuring to know others have been there and made it through. ”

– Kate, Mum of Alice
aged 3.5yrs 2012

“ The liver transplant journey is different for everyone – but 'potholes' and really rough days are guaranteed. Putting one foot in front of the other, remembering to focus on the positives, to cry when I felt like and to talk when needed was what helped to get me through the worst of it. 20 months on from transplant Isabelle is a cheeky, chatty two and a half year old who attends pre-school three days a week, and as her live donor Mum I am now excited about baby number two arriving in a few months. ”

– Jodie



Heroes



*Te Pare (Utiku's Aunty) with
Gina mother of Utiku*



TK with his donor mum TP



Shania with her Mum

A Recipient's Perspective...Poem

“ Discomforts of needles, staples, and stitches soon became my friend,
knowing now this would no longer be my end.

My disease had certainly taken its toll,
destroying my liver, but never touching my soul.
With love and support from my family and friends,
I learned not to break, but to accept and to bend.

It has been 16 years now from that day to this,
No need to ponder an after-life filled either with pain or bliss.

I'm alive and I'm well and on a list no longer wait.
Now, I've more energy and extra time to create.
There's a sense of achievement I seek to fulfil. ”

“ My mumshe gave me a second life.
MUM means the world to me. ”

“ I am an active
sports person.
I was also awarded
a sportswoman of
the year. ”



*Shania born in Fiji, transplanted in New York via
compassionate funding, living
in New Zealand*

Karama's Journey with Biliary Atresia

by Tala

There are some memories in your lifetime that you will always remember vividly, right down to what you were wearing on that day. That holds true to when we were told of Karama's diagnosis. I don't remember everything that was said but I do remember these words, rare liver disease, biliary atresia, kasai, and possible transplant required. I had heard enough. The tears started to fall and there was a rustle amongst the gastro team who had all crowded into our tiny room and I knew they were trying to find me tissues which for some reason made me feel bad. I remember Karyn coming in after the team had left to explain further what Biliary Atresia was. I don't think I heard a word she said, I was still numb and don't think I cared, the only thing I had in my head was he was seriously sick.

Following the kasai, we went home. We were very optimistic that Karama would be the exception as we were well aware of the odds of the kasai actually 'fixing' him. Alas, this was not the case. Our next trip back to Starship was to do the work up for transplant. The word transplant had always been frightening, however in the short time that we were home with him, he had started to deteriorate and our lives were in limbo. So we now looked forward somewhat to having him transplanted as this would 'sort' it. Hopefully.

We spent the week in Starship meeting all the specialists who would be involved with his care prior and following transplant. We were well informed and had a better understanding of what was to come. The handbook we were given was

useful as it listed signs to look out for and complications associated with the disease. As the disease progressed, I found myself ticking off all the complications that was listed in the handbook. I remember initially being scared if he happened to get one, but for him to get all of them, well nothing could prepare me for that.



Our lifeline was knowing that he could also be transplanted via live donor. Sorted, his dad was in good health and should not have a problem with BMI, so we would move on and get him transplanted. After his dad went through the vigorous testing, we waited for what seemed like forever only to be told that his dad was not compatible. Back to square one.

Months passed and I naively thought, he'll get one, a deceased liver will come up soon and it'll be his. I did not take into account that he would catch pneumonia several times which meant he was suspended off the list for a week. Those weeks sucked, all we were doing was waiting, and to have a week go by where had a suitable liver come up and we could not even be considered for it, was absolutely heartbreaking. The waiting game truly sucked and I did not want

to play it anymore.

Christmas was coming and I was dead set on him having been transplanted by Christmas, surely he had waited long enough and this would be his ultimate Christmas gift. Again, I was wrong. The New Year came and went, I was only grateful to say goodbye to 2011 as it was the worst year of our lives.

January 10th 2012, Karama was transferred to PICU. It hadn't been his first visit to PICU so I assumed it would be another few days there to be 'stabilised' and then we'd be back up to the wards to wait. Again I was wrong. He had deteriorated quickly and this was the worst I had seen him. For the first time, since he was diagnosed, I was scared. Scared that he would not be travelling home with me after all. I remember his dad had said to me that morning that I needed to be 'ready'. I knew what he meant, but to hear him say it out loud, I could have literally throttled him with my bare hands. Somehow Karama got through another day. But my mindset had changed. I had sadly lost hope that he would get a donor in time, so I sat by his bedside again, waiting. But this time I was waiting for his little body to finally give up. Karama from





the start had taken the lead and we had no choice but to follow. Looking at him lying there, stomach bloated, supported by ventilator, I felt that he was telling us he was tired and wanted to 'rest'.

And then it happened. James the anaesthetist aka angel of good news, walked in and told me what I'd been waiting to hear. A possible match had been found in Australia but this would not be confirmed until around lunchtime. He then proceeded to tell me he was going to start putting lines and so forth in just in case it went ahead and that I should take a break and he'd ring me once he was done. I understood and heard every word he said clearly, but out of my mouth came, 'what do you mean?' To finally hear what we'd been waiting forever to hear was just numbing. That day was the longest day ever. Finally it was confirmed the deceased liver was good and on its way. As he was wheeled into theatre, it was still surreal

that it was truly happening.

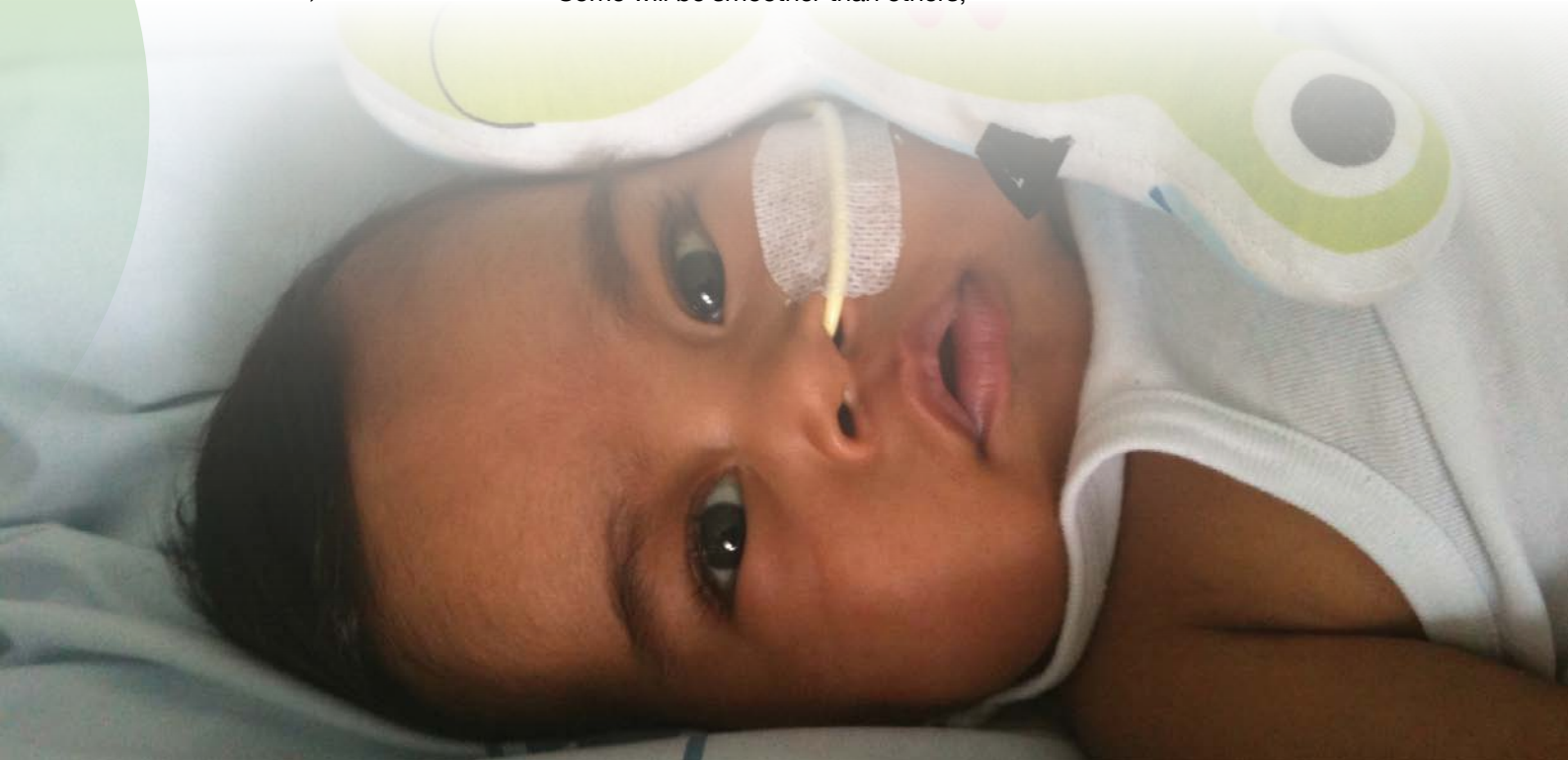
Karama's recovery post-transplant was slow and not without complications. Story of his life really. But again, we followed his lead and slowly but surely, he started to improve. One by one wires and lines started coming off, all except his NG tube remained. As each day went by, he started to look and feel like a 'normal' baby. It hasn't been a smooth road to recovery but I would not expect anything less from him. He has tested us beyond our limits, and then some. But we were discharged home just in time to celebrate his 1st birthday the next day.

Today we are almost 4 months post-transplant. I have nothing nice to say about the disease. In one word? Shit! That alone doesn't come close to summing up our experience. What I do know is that every child with BA, although the disease is the same, each child although they travel the same path, all the paths will be different. Some will be smoother than others,

where as ours was definitely laden with potholes, sinkholes and every other kind of hole that he could fall into, he dove right on in.

Nevertheless, the boy I look at today, with his big brown eyes, and beautiful eye whites that once were yellow, is amazing. He smiles as though he's had it easy in his short year of life. To see him smile today and play with his brothers, well it just makes the turmoil he'd put us through, fade into the background.

Still have to get through his 'bubble' year, but his future is looking brighter already. Our biggest accomplishment is being blessed with this child. He has reminded us how precious a gift life truly is and that we shall no longer take it for granted.



Writing to the families of organ and tissue donors



A tribute at the Transplant Games in Brisbane, to acknowledge donors.

The decision to write is a personal one.

It can be difficult to find adequate words to convey your gratitude.

Many organ and tissue recipients would like to communicate their thanks to the donor family and share a bit about themselves following transplantation. It can be difficult to find adequate words to convey your gratitude but it is important to remember that this correspondence will be hugely appreciated by most donor families.

Often it can be daunting to write a letter to the donor family – a card might be a better option for you. You are encouraged to wait at least three months after the transplant before writing. This gives your child and the donor family time to recover.

Please remember that your transplant team and Organ Donation New Zealand (ODNZ) will not facilitate meetings between transplant recipients and donor family members. Confidentiality is maintained to protect the privacy of everyone involved.

A few tips for writing to the donor family

- You may wish to say thank you to the donor family and recognise the death of their family member. Talk about your child and your family.
- It might be helpful for the donor family to hear how the transplant has changed the life of your child and that of your family. Use language that is respectful and sensitive. Remember that the family are coming to terms with the death of a loved one.

- Don't include identifying information such as your name, where you live, where you work and phone numbers.

All correspondence is reviewed and nothing will be sent that contains this information.

- Simply sign the card with "from a grateful recipient family" or something similar.

When you have finished writing:

1. Place the letter or card in an unsealed envelope
2. On a separate piece of paper write your full name and the date of your child's transplant.
3. Send both to your Transplant Coordinator/CNS

Your message will be forwarded to the donor family via your transplant team and ODNZ . The donor family may indicate that they are not ready to receive your correspondence.

If this is the case your letter will be held by ODNZ and forwarded when the family is ready. Your transplant team will let you know if your correspondence has not been passed on to the family.

It is also important to remember that while some donor families may choose to respond to your correspondence, some may not. Please do not take this personally. Some donor families may respond years later.

Sharing your story

Sometimes recipients like to share their personal transplant experience. If you are speaking to the media or sharing your story with the general public, please remember not to use exact dates. The same applies if you have a presence on the internet for example a Facebook profile, blog or other social networking sites.

If you have any questions or concerns, please contact the CNS / Transplant Coordinator.

Terminology

Abdomen Part of the body which contains the stomach, liver, spleen, kidneys and pancreas.

Antibodies A Substance, which is produced in response to specific antigens, which help fight infection and foreign substances.

Antigens A substance, which triggers an immune response.

Ascites Fluid which accumulates in the abdomen.

Bile A yellow–green fluid made in the liver and stored in the gall bladder, which breaks down fat.

Biliary System The network of bile ducts inside and outside the liver.

Cholangitis Inflammation of the bile ducts.

Cholesterol A form of fat that is found in the body, which performs necessary functions.

Cirrhosis Widespread hardening and scarring in the liver causing obstruction of the flow of blood.

Creatinine A substance found in blood and urine. A high blood creatinine level indicates a reduction in kidney function.

Haematemesis Vomiting of blood which can be fresh (bright red) or old (dark, looks like coffee-ground particles).

Hepatomegaly Enlargement of the liver.

Hypertension High blood pressure.

Jaundice A Yellow discoloration of the skin, whites of eyes and mucous membranes.

Liver Enzymes Substances produced by the liver and released into the blood. A Rise in enzymes may indicate rejection.

Melaena The presence of blood in stools. This can be black (old blood) or dark red (active bleeding).

Oedema Excess fluid in the tissues, usually around the ankles, legs, hands and fingers.

Rejection A Complication of transplantation, where the body recognises the new liver as a foreign body and attacks it.

Splenomegaly Enlargement of the spleen.

Strictures Narrowing of blood vessels or bile ducts.

Variceal Bleed Bleeding from varicose veins within the oesophagus or stomach. A complication of chronic liver disease.

Varices Varicose veins of the oesophagus or stomach.

Sample letter for schools and kindergarten

Department of Paediatric Gastroenterology
Room 5.159
Starship Children's Health
Private Bag 92024
Auckland

Team Support: Dani Ta'ase Ext 5471

Attention: Principal and class teachers for Lara

Dear Sir/Madam

Lara has previously had a liver transplant and will be commencing at your school shortly. We expect that Lara will be able to fully participate in the school community both in class and in the playground and should be given every opportunity to participate. Some children after having had a major illness in infancy can experience learning difficulties. Please have a low threshold for referral to specialist services. But there are a few considerations you will need to take into account when dealing with Lara. The first is that Lara is on medications to prevent her rejecting her new liver and these work by suppressing the body's immune system. This makes Lara more susceptible to contracting infections such as colds and impetigo.

Good hygiene habits are an important part of infection so if Lara and her classmates can be encouraged to remember hand washing after the toilet and before food where possible that would help greatly. Some schools have implemented the use of hand gels by doors for use as children enter and exit the classroom as a means of addressing this issue.

Infectious diseases such as Chicken Pox, Measles, Impetigo and Gastroenteritis are of concern in children with low immune systems and may require special care. Therefore we ask that the school maintain an awareness of these occurring in the school and alert the family to the presence early so they can make decisions regarding appropriate action. Please provide the parents with information regarding the potential contact between children for example; is the unwell child in the same class or have been known to play directly together as this helps in the decision making process for further treatment.

Should Lara become unwell during the school day please contact the parents at the earliest opportunity. In emergency situations please call an ambulance as per your normal process and tell them this person has had a liver transplant.

We encourage children post transplant to be involved in sport but contact sport such as rugby, martial arts etc are not appropriate as the new liver often remains lower than a normal one and may not have the normal protection of the rib cage.

Lara should not need to have access to transplant medications during a normal school day as they are given morning and night. However please liaise with parents as this will need to be addressed for school camps and any long day outings. We currently recommend that a small supply (a few days worth of medication) should be kept at school for a Civil Defence Emergency situation where she may need to remain at school overnight or longer, as it is important that Lara does not miss doses. The family will discuss appropriate arrangements which suit your school.

Please do not hesitate to contact us should you have any further questions or concerns.

Kind regards

Karyn Sanson/ Cate Fraser-Irwin
Paediatric Liver/Gastro Nurse Specialist Service
Phone: 09 3074949 Extn 29085 / 29043

Important telephone numbers

Starship (Auckland City Hospital)	09 307 4949
Liver Nurse Specialist Cate Fraser-Irwin	09 307 4949 Extn 29043 021 660 299
Karyn Sanson	09 307 4949 Extn 29085 021 938 854
Gastroenterology Consultants	Care of Dani (Team Support) Extn 5471 Out of business hours please contact On-call Consultant via Auckland City Hospital switchboard 09 307 4949
Gastroenterology Fellow	021 613 729
Ward 26B Medical Specialties	09 307 4949 Extn 25760
PICU	09 307 4903
Social Worker	09 307 4949 0212416268
Pharmacist	09 307 4949 Extn Pager No 935059 / 937471
Dietitian	09 307 4949 Extn 7248 Pager No 935010
Ronald Macdonald House	09 303 1365
KIDS Foundation	0508 300 600 (Free phone) Ask for the support coordinator when you are in Starship. http://www.idfnz.org.nz/

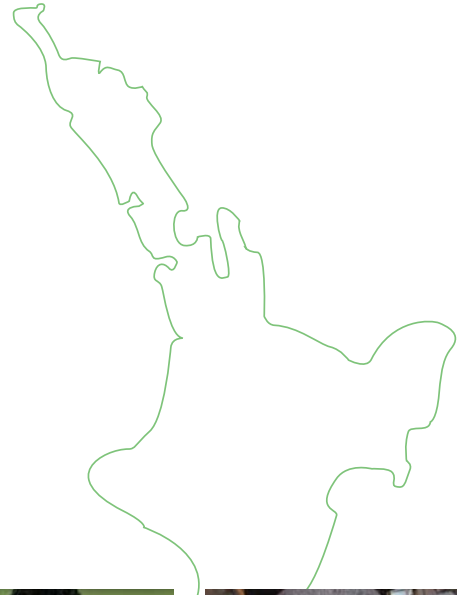
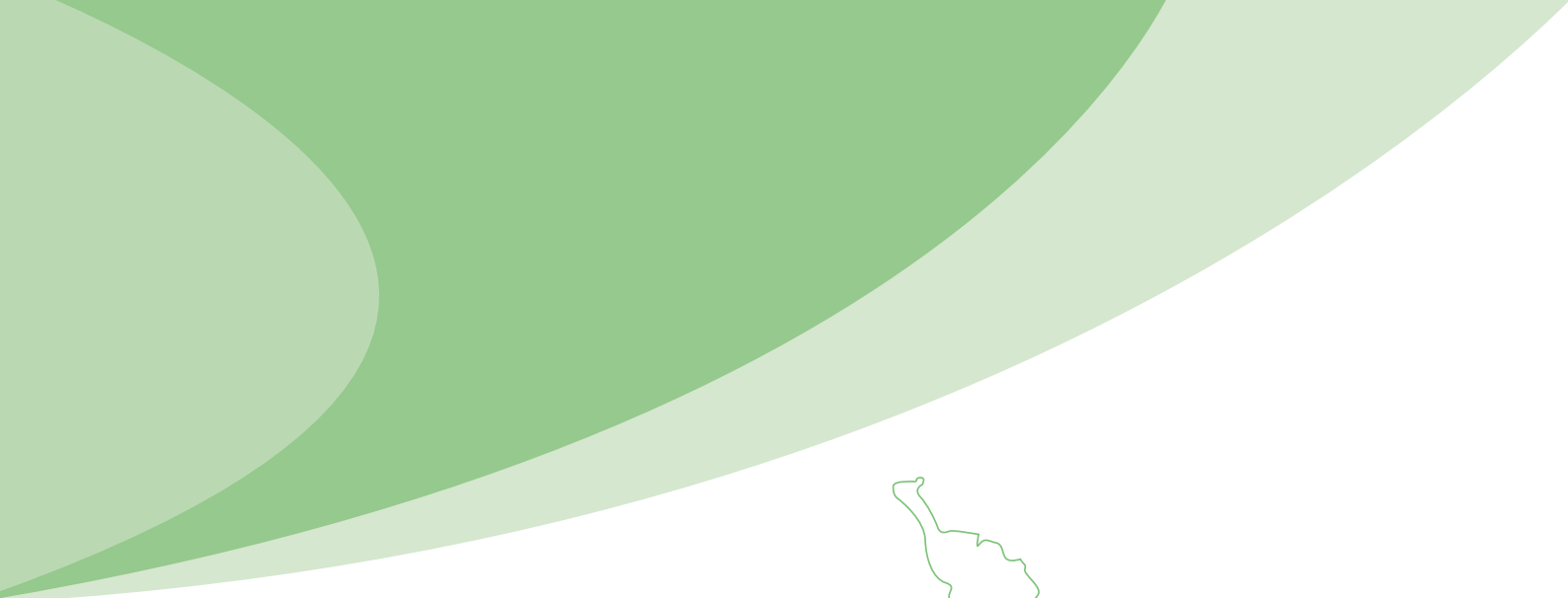
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Notes

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The
Kids
Foundation
key immune deficiency support