

Intrathecal baclofen (ITB)



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Your ITB team

Your ITB nurse is: _____ Telephone: _____

Your ITB doctor is: _____

Your managing ITB team is: _____

Business hours please contact: _____

After hours please contact: _____

Notes: _____

This book was written by members of the Complex Movement Disorders team, The Royal Children's Hospital, Melbourne. We gratefully acknowledge the generous financial support of the Waverley Auxiliary who provided the funding for printing this booklet.

Introduction

What is intrathecal baclofen?

Intrathecal baclofen (ITB) is a well established therapy for children with movement problems. Baclofen is a medicine that helps the muscles relax. It can be given by mouth or 'intrathecally' which means delivered into the space around the spinal cord. Oral baclofen is usually tried first but if it doesn't work well, intrathecal baclofen might be considered.

ITB is injected into the fluid around the spinal cord through a very fine tube (called a catheter). This tube is connected to a surgically implanted permanent pump that is placed in the abdomen.

This booklet contains general information to help parents and families understand the ITB treatment process, and describes services available. Further information is available on The Royal Children's Hospital website at www.rch.org.au/kidsinfo and useful contact details are printed on the back cover of this booklet.

How you say it:

Intrathecal *In-tra-thee-cal*

Baclofen *Back-low-fen*



Who might be suitable for ITB?

Children and adolescents with moderate to severe spasticity or dystonia that affects care, comfort, quality of life, and function may benefit from ITB. Many of these children and adolescents have cerebral palsy; many others have an acquired brain injury (ABI) following trauma or stroke, or other more rare neurological conditions. However there are many factors to consider when deciding if your child should have ITB treatment. ITB does not suit every child or every family. Being part of the ITB program puts limitations on certain parts of your child's life. You have to live close to a medical centre that can manage the ITB and you have to be able to attend many appointments. All the people that care for your child need to be willing to learn about ITB and its side effects; your child's age and weight also need to be taken into consideration.



What is spasticity?

Spasticity is a motor disorder where the muscles contract involuntarily making the muscles tighter than they should be. It is usually caused by damage to the portion of the brain or spinal cord that controls voluntary movement. The degree of spasticity (or 'stiffness') can vary from person to person. It may occur in children with cerebral palsy, ABI, and other more unusual conditions of the brain or spinal cord.

What is dystonia?

Dystonia is a movement disorder where there are sustained muscle contractions that frequently cause twisting or repetitive movements or abnormal postures. These movements can be painful and cannot be controlled by the person. Dystonia can affect just a single body part or be generalised to most or all of the body. Dystonia can also result from the same causes as spasticity, that is, cerebral palsy, ABI and other more unusual conditions of the brain and spinal cord. It may also occur as a result of an adverse drug reaction or a brain tumour.

Complex Movement Disorder program

Referral, consultation and assessment

A referral to the Complex Movement Disorder (CMD) program can be made by members of the CMD team, or other doctors or therapists from within the hospital. Once a referral has been made for assessment for ITB, your child's case history will be presented at a CMD team meeting.

At this meeting your child's suitability for ITB will be considered. The CMD team will review your child's medical history, past treatments, video footage of your child's movement problem (if available), and make an assessment of whether ITB may be appropriate. At this team meeting, other forms of therapy are also considered.

The first stage of assessment usually involves a consultation with either the paediatrician or rehabilitation physician. This is to ensure that other forms of treatment, such as oral medications for spasticity or dystonia have been tried, and also to ensure that other medical problems, which may make spasticity or dystonia worse have been treated.

If thought that your child may be suitable for ITB, your child will then be assessed in the CMD clinic.

The CMD program at The Royal Children's Hospital (RCH) comprises a multidisciplinary team made up of doctors, therapists and nurses.



COMPLEX MOVEMENT DISORDER TEAM MEETING

Who is in the CMD team?

- Cerebral palsy clinical nurse coordinator
- Clinical nurse consultant – intrathecal baclofen and complex movement disorders
- Consultant paediatrician/rehabilitation physician
- Gait laboratory physiotherapist
- Neurologist
- Neurosurgeon
- Occupational therapist
- Orthopaedic surgeon
- Physiotherapist
- Speech pathologist
- Rehabilitation nurse coordinator

CMD assessments

ITB assessment team

The CMD clinic is a multidisciplinary clinic. Your child will be assessed by a number of different health professionals working together. The assessment takes about one and a half hours. The assessment team includes:

- Consultant paediatrician/rehabilitation physician
- Neurosurgeon
- Clinical nurse consultant – intrathecal baclofen/complex movement disorders
- Physiotherapist
- Occupational therapist
- Speech pathologist

What to expect

At the CMD clinic, you and your child will be asked a series of questions. These will focus on your child's day-to-day life, and their abilities and challenges. An important part of the assessment is to understand what you and your child hope to achieve with ITB, and on the basis of this, to set some clear and specific goals.

Your child's community or school-based therapist may also be contacted prior to the clinic assessment. They are invited to attend the assessment. In their absence we ask them to complete a written assessment of your child's movement disorder, current issues and priorities.

Physical assessments

Your child will also have a series of physical assessments. These assessments range from a physical examination, to your child carrying out specific tasks. The assessments will vary depending on your child, and goals of treatment. These assessments are video-taped. These videos are only used for the assessment and follow-up of your child in the CMD program. The videos can only be used with the specific permission of you and your child. Your child's equipment needs and therapy requirements will also be discussed.

At the end of your appointment

Following the assessments, you will be given more detailed information about ITB. You will also be given written material to take away with you to read.

You and your child will have a chance to ask questions. It might be helpful to write down any questions you may have, before the visit.

After your child's CMD assessment

You will be given an opportunity to think about everything that has happened at the CMD clinic, and about the information you have been given. You are encouraged to talk with your family and other support people about ITB. All the results of the assessment are then discussed with the CMD program members.

Together the team decide whether your child should go to the next step of the process which involves a trial of ITB. You will be contacted after this discussion to see if you have any other questions, and if the team recommend that your child may be suitable for a trial with ITB you will be asked whether you would like to proceed with the trial.

ITB therapy trials

The next stage of the assessment is a trial of ITB. The aim of this stage is assess whether the ITB works for your child before implanting an ITB pump.

There are two types of trial:

1. Bolus trial
2. Catheter trial

The type of trial will be determined at the assessment.

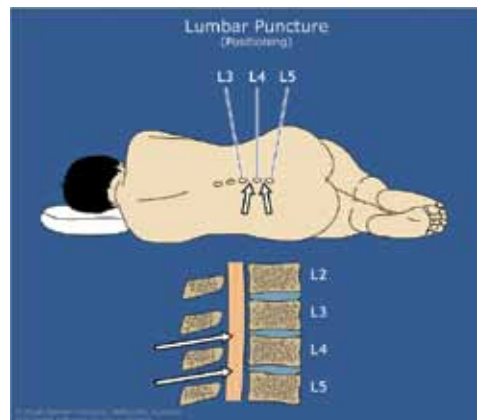
1. Bolus trial

A bolus trial involves the injection of a single dose of ITB into the space surrounding your child's spinal cord. The bolus is given via a lumbar puncture. A lumbar puncture involves introducing a small needle into the space surrounding the spinal cord. The ITB is then injected through the needle. The lumbar puncture will be performed either under sedation from nitrous oxide (laughing gas) or under general anaesthetic. This procedure takes 30 minutes or more.

Your child will be admitted to a ward and must remain in bed for at least two hours after the lumbar puncture. A doctor, physiotherapist and occupational therapist will observe your child approximately every two hours and assess how baclofen has affected your child's muscles. The therapists will examine the stiffness and range of motion in your child's muscles. The ITB usually starts to work about one hour after the injection. The effects of baclofen start to wear off after four hours.

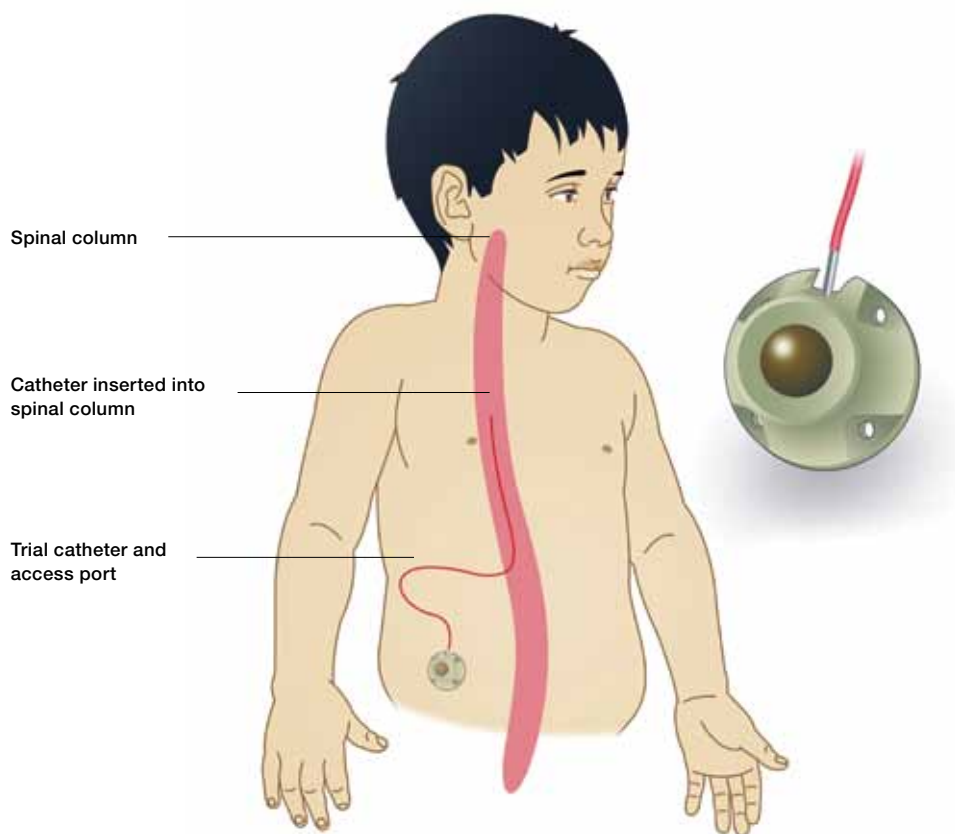
The lumbar puncture may cause your child to develop a headache, and he/she may also experience side effects from the anaesthetics such as vomiting. If this occurs, your child may have to stay overnight for further observation. However if your child is well, he/she will be discharged in the late afternoon.

If the trial is successful and you and your child wish to proceed and have a pump implanted, an appointment will be made for you to see a neurosurgeon to discuss the option of a pump and the procedure involved.



LUMBAR PUNCTURE PROCEDURE

POSITION OF THE TRIAL CATHETER AND ACCESS PORT



2. Catheter (tube) trial

The catheter trial is more extensive than the bolus trial. It allows assessment of what effect higher doses of ITB will have on your child's movements. For this type of trial your child is required to be admitted for two inpatient stays.

Staying in hospital for the catheter trial

Stay 1. Insertion of the catheter and access port

The catheter and access port are put in by the neurosurgeon whilst your child is in the operating room and is under a general anaesthetic. The insertion of the port and catheter is usually a separate admission to the trial. The catheter is inserted into the spine and is then connected to an access port, which sits just under the skin on the abdomen.

Your child will then be discharged home to recover from this surgery. He/she will be readmitted at a later date for trial of the ITB. Your child may have some pain resulting from the surgery. Regular analgesia (pain relief medication) will be given and staff will ensure that your child remains comfortable.

Stay 2. Trial of ITB

Your child is readmitted for an inpatient stay of approximately 3–5 days. This is usually within four months of the port being implanted. A needle is inserted through the skin into the access port. A local anaesthetic cream called 'Angel' or 'Emla' cream is placed on the skin over the access port site on the abdomen. This cream will numb the skin so there should be no pain from the needle.

The needle is attached to tubing and a small external pump. The baclofen will be delivered to the spine by the pump.

Only your baclofen doctor and the clinical nurse consultant will alter the dose of baclofen.

During the trial your child will be able to get out of bed and move around – that is walking if they are able, or moving around in their wheelchair. Throughout the trial your child will be assessed for the effect baclofen has had on your child's muscles. The therapists will examine the stiffness and range of motion in your child's muscles. In addition, your child will be assessed and videotaped performing specific activity goals that he/she would like to achieve with ITB treatment. During the trial the amount of baclofen may be altered to assess the impact of the altered dose on your child's movements while monitoring for any side effects.

When all the assessments are complete and the ITB team are satisfied with assessments, the medication is stopped and the needle is removed. Your child will be discharged home.

If an improvement is observed in your child's muscle tone and you wish to proceed, a date will be made for the access port to be removed and the baclofen pump will then be attached to the catheter that is in place. This may be done during the same admission or you may need to return to the hospital at a later date.

Useful information about RCH

For more information about RCH and its facilities, please read 'Your Guide to The Royal Children's Hospital'. This is a free booklet and is given to families at the time of admission. It is also available in all clinic areas and the Family Resource Centre at RCH. It is also available online at:

www.rch.org.au/rchinfo

Operation for insertion of ITB pump

Before the operation

The night prior to your child's operation, you will need to wash your child's skin with an antibacterial soap wash called triclosan, to help reduce the risk of infection. This is available at your local pharmacy. When the date of your child's operation has been decided you will receive a letter from the department of Neurosurgery. This letter will tell you when and where you and your child need to be on the day of surgery. This letter will also tell you when your child needs to fast (stop eating). If you have any questions about the information in this letter please call your ITB nurse.

You will need to bring personal items such as comfortable pyjamas and toiletries for a 3–7 day stay. Please bring your child's favourite/comfort toy to help settle and calm them whilst in hospital. Please ensure all your belongings are labelled and leave all unnecessary valuables at home.

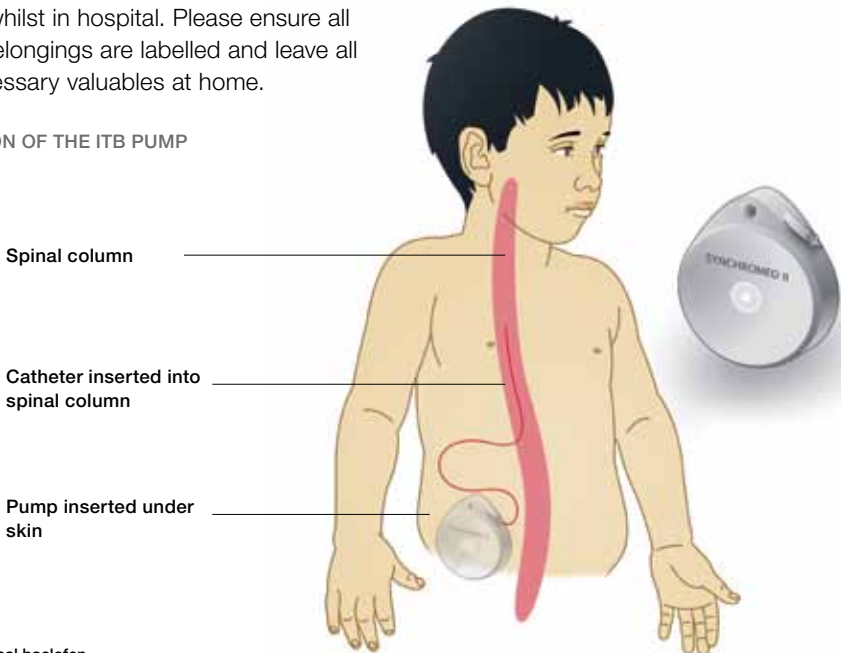
During the operation

You are able to stay with your child in the operating room until the anaesthetic has started to work. The operation to implant a pump takes approximately two hours.

If your child has had a bolus trial, a catheter is placed in the spinal column (an intrathecal catheter). The end of this catheter can sit as high as the shoulder blades or as low as the waistline depending on what type of movement condition your child has.

Once the catheter is in place, the pump is attached to the catheter. If your child had a catheter trial, the catheter is left in place. The surgeon removes the port from the end of the catheter and attaches the ITB pump.

POSITION OF THE ITB PUMP



Following the operation

Your child will be observed in the Recovery area straight after the operation. When he/she is awake enough they will be transferred to the Children's Neuroscience Centre on the 8th floor of the hospital.

Children need to stay lying flat in bed for the initial 48 hours after the operation. Your nurse will ensure that your child is comfortable and has adequate pain relief medication.

Your child may have a urinary catheter (tube) inserted into their bladder, when they are in theatre. This catheter allows urine to drain into a bag. This prevents urine leaking out of nappies or continence pads and contaminating the surgical wound. It is very important that the wounds and dressings are kept dry to reduce the risk of infection. Usually the catheter is removed as soon as your child is able to get out of bed.

Within two days of surgery your child will have an x-ray of their spine. This x-ray is to check the position of the catheter.

Staying in hospital

Your child is likely to be in hospital for 5–7 days to recover from this surgery. You will see the ITB team often during your child's stay in hospital – sometimes more than once a day. They will gradually increase the dose of ITB.

During this time your child will also be reviewed by the physiotherapist. This is particularly important if your child is in a wheelchair, as adjustments may need to

be made to the chair to accommodate the ITB pump. In addition your child may need a protective dressing, or soft belt, to protect the pump from pressure.

Baclofen can slow down the muscles in the bowel which can cause constipation. If constipation is a problem for your child, please let your ITB doctor know and they will treat it with medication.

The dose and concentration of ITB that your child receives will determine when he/she is due for their first pump refill. The ITB team will book an ITB appointment before your child is discharged.





Follow-up after your child's stay

Your child will have regular, often weekly ITB appointments at the hospital after discharge, to gradually increase the intrathecal baclofen dose to the right level for your child. During these appointments the surgical wounds will also be observed. You will also have a follow-up appointment with your neurosurgeon.

At 3, 6 and 12 months after the pump has been implanted arrangements will be made for a physiotherapist and occupational therapist to review your child to assess:

- what effect the ITB has had on your child's muscles
- if your child needs additional occupational therapy or physiotherapy to assist with the changes the ITB has made.

A report of the assessment findings will be sent to your child's local therapists. This report may include recommendations for changes in your child's current therapy program.

Your child's local therapists, carers, GP, staff at school or paediatrician may have questions about ITB. Please encourage them to contact the ITB nurse consultant for further information about ITB.

When your child has an ITB pump

Refills

The ITB pump is refilled every 1–6 months depending on your child's dose.

During the first six months your child will be required to come in more frequently to check the baclofen effect, alter the dose as needed and to monitor how your child is progressing.

Pump refills are performed during an outpatient appointment at RCH. Your child does not have to be admitted to hospital for this procedure.

The skin over the pump is numbed using an anaesthetic cream called 'Emla' or 'Angel' cream. It takes approximately 30–45 minutes to work, so it is important to put the cream on prior to coming to your appointment. You will be shown how to do this before your first refill.

Usually, after the 'Emla' or 'Angel' cream has taken effect, your child should not feel the pain from the needle. Some children describe feeling the pressure of the needle going in.

The pump refill procedure

The ITB pump refill is a sterile procedure. It is very important to avoid infection.

The paediatrician, rehabilitation physician or clinical nurse from the ITB team performs the procedure. The area around and over the pump site is thoroughly washed and cleaned with chlorhexidine (antiseptic alcohol). A sterile drape is then placed over the pump and the abdomen to create a sterile working area. It is very important that no one touches the drape, to keep the area free of germs. A needle is inserted through the skin and into an access port in the middle of the surface of the ITB pump. Once any remaining baclofen has been removed, the new baclofen is injected into the pump. The pump is then reprogrammed and the next refill date is calculated. Dose changes can also be made at this time.

REFILLING THE ITB PUMP



Programming the pump

The ITB pump can be programmed a number of ways to suit individual needs. The pump can deliver a steady dose of ITB over a 24-hour period. Boluses can also be programmed at different times of the day.

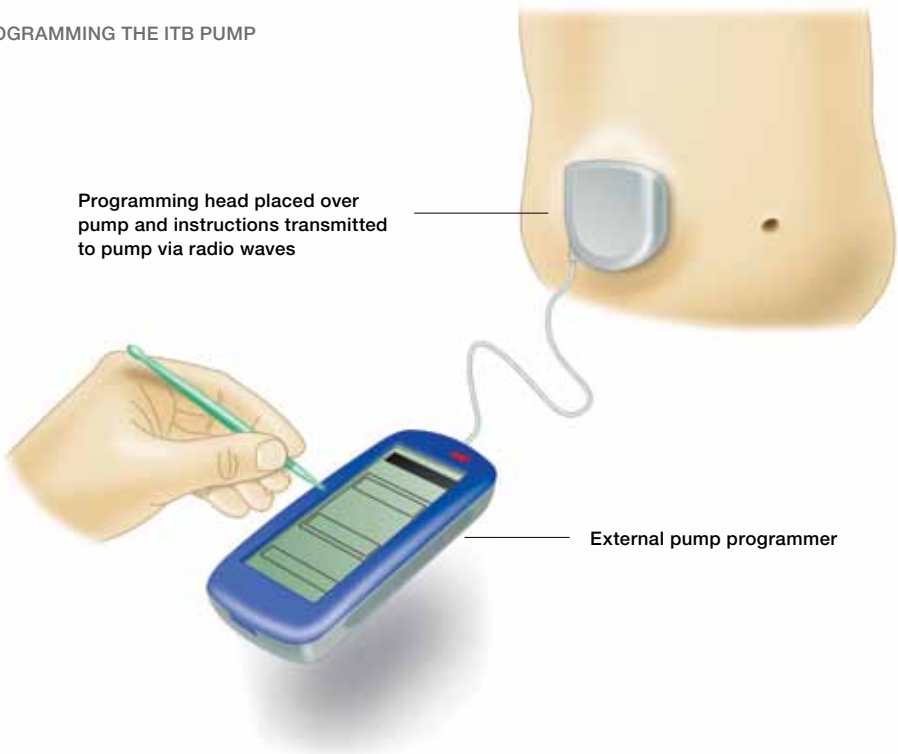
Activities

It is important to note that once the wound has healed, your child can participate in all of his/her usual activities, perhaps better than ever before, given the significant benefits that the ITB pump will provide. However care should be taken and the pump should be protected. Your neurosurgeon will tell you when it is safe to resume normal activities.

Holidays

When planning a holiday, you must always consider how you will get help if you have concerns about your child's pump. Will you have access to medical help that have knowledge of the pump? If not, who will you contact, where is the nearest hospital? What is the emergency number (if travelling to another country)? If you are planning a holiday, talk to the ITB nurse, who will be able to help you.

PROGRAMMING THE ITB PUMP



Complications

There are some important things to be aware of if your child has an ITB pump. Complications are rare but it is important to know the side effects and what to do if something goes wrong. If an ITB pump or catheter malfunction is suspected your child will be readmitted for investigation. If you are concerned about your child, you can contact the ITB team at the RCH. Refer to contact details on the inside of the front cover of this booklet.

Possible complications of ITB therapy:

- Overdose of baclofen
- Under dose of baclofen
- Infection of pump or catheter
- Infection of wound
- Catheter kink (i.e. bent tubing), preventing the flow of baclofen.
- Catheter break – causing leak of baclofen
- Catheter disconnection from pump – preventing baclofen from reaching spinal cord
- Pump malfunction (pump not working properly)

When to call your ITB doctor

- If your child has a temperature higher than 38°C
- If you notice any redness or inflammation around the scar
- If the scar changes (i.e. changes colour, has ooze or bruising)
- If there is any swelling around the pump area
- If your child is more sleepy than usual
- If you think your child is too 'floppy'
- If your child is unusually sweaty or itchy or grumpy
- If you are concerned about the pump
- If you hear beeping from the pump
- If you think your child is receiving too much or not enough baclofen (see side effects on page 14)

An ITB doctor is on-call 24 hours a day, 7 days a week. Please see the contact details on the inside of the front cover of this booklet.

Side effects of baclofen

Baclofen can have side effects. These include:

- Constipation (can't do poos easily and poos are hard)
- Nausea (feeling sick)
- Blurred vision
- Decreased tone (beyond what is desired)
- Depression
- Headache

Not enough baclofen

It is important to know that after a few months your child's body will become used to baclofen and if it was to stop suddenly they might experience some symptoms. Signs that your child may not be receiving baclofen:

- Your child's muscles are tighter than usual
- They have a high temperature
- They look flushed and hot
- Their skin gets itchy
- They are grumpy or irritable

If, for any reason, they suddenly stop getting baclofen it can make them very sick.

Too much baclofen

It is also important to know that sometimes children get too much baclofen. This may happen when a catheter is kinked and then becomes unkinked, or from human error when programming the pump, or because your child is sensitive to an increase in their dose of baclofen. It is important to know the signs and symptoms of too much baclofen.

Signs that your child might be getting too much baclofen:

- Drowsiness
- Their body is more floppy than usual

If you think your child is not getting enough baclofen, or if you think they are getting too much you **must** take them to the emergency department at RCH and contact the ITB team — call (03) 9345 5522 ask for the Developmental Medicine Consultant on call.

Remember you can call 000 for an ambulance at any time if you are concerned.

Call an ambulance immediately if:

- Your child has slow breathing
- You can't wake your child

Pump beeping

The pump is designed to beep if the medicine is running out or if the battery is going flat. This is a warning beep.

The battery life of a pump is approximately 5–7 years. The pump will begin to beep if the battery starts to run out before the pump is replaced. Before the battery becomes flat your child will have another operation to put in a new pump.

If, in unusual circumstances, your pump is not refilled when it is due, the pump will 'beep' two days before the medicine is due to run out.

What to do

If you ever hear beeping from your child's pump, even if you think it is not due for a refill you MUST contact the ITB doctor and take your child to the RCH Emergency department. See contact details at the back of this booklet.

Tell doctors that your child has an ITB pump

Every time your child is in any hospital for any reason you **must** tell the doctors that your child has an ITB pump. Ask the medical team to contact the RCH ITB team as per the contact details on the back cover of this booklet.

Make it a habit to tell every medical person (doctors, nurses, radiologists, pharmacists) caring for your child that they have an ITB pump. This way they are aware of the pump and will consider it whilst caring for your child.

MRIs

If your child requires an MRI, it is important to let the RCH ITB team and the medical staff at MRI know that your child has an ITB pump prior to your MRI. MRIs can affect the pump's mechanics. Your child can have a MRI, but once the MRI is completed, their ITB pump will need to be reprogrammed. The team is able to organise for someone to be present at the MRI to reprogram the ITB pump. Tell all medical staff that your child has an ITB pump.

Key points to remember

- Complications from an ITB pump are rare but you must always be alert and contact your ITB doctor if you are concerned.
- If you hear beeping from the pump, contact your ITB doctor urgently or bring your child to RCH Emergency department.
- You can always call an ambulance 000 if you are worried about your child, even if you are unsure.
- If you are unsure of any of this information, call the baclofen clinical nurse consultant. See the inside front cover of this booklet for contact details.



Frequently asked questions

Can the catheter break?

The catheter is made of a very durable and stretchy plastic called silastic. It is very rare for a catheter to break, but it does happen. If a catheter breaks, the baclofen would leak and your child would not receive their full dose. They may then experience the symptoms of withdrawal mentioned earlier in the booklet. There are scans and tests that can be done to see if the catheter is broken. If it breaks, then your child will have to have an operation to have a new catheter put in place.

How long will the pump last?

The battery on the pump lasts about 5–7 years. At this time your child will need another operation to replace the pump.

How frequent are refills?

ITB medicine refills will be every 1-6 months depending on the dose your child needs. The first refill may not be for several months after the operation but may be more often after that time. This is because your child will start on a very small dose and it will take longer for the pump to run out.

How much baclofen will my child be given?

This depends on your child and how they respond to the baclofen. Your doctor will discuss all dose changes with you before any changes are made.

Can my child still go swimming, have spas and baths once the pump is implanted?

Your child can usually go swimming and have baths six weeks after the pump is implanted. This is as long as the wound

has healed and the neurosurgeon has given the 'OK' to do so.

Can the pump be exposed to very hot or very cold temperatures (such as a hot water bottle or an ice pack)?

The pump is designed to tolerate a wide range of temperatures. It is fine to use both hot and cold packs near the pump. Spa baths are also fine but saunas should be avoided.

Can my child play sport with the pump in?

Your child should not play high velocity contact sports, as the pump may be knocked out of place. Non-contact and wheelchair sports are fine. Discuss this with your ITB doctor, as you may need to get some protective padding to cover the area around the pump to cushion any blows to the area.

What happens at the airport at security check-in?

The pump is made of metal and will set the alarms off. You can ask your ITB doctor for a letter to explain the pump. The scanners will not affect the way the pump works.

Will the pump sticking out cause any problems?

Some patients say they needed to buy looser clothes (underwear, trousers) as normal fitting clothes can cause discomfort over the pump. Before you leave hospital, your physiotherapist and occupational therapist will assess the pump area and how it fits into your child's wheelchair (if they have one). If needed, your therapist can give you some padding to cushion the area, and/or cover and protect the pump.

Contact information

Emergency

(Ambulance and Emergency Services)

Telephone: 000

The Royal Children's Hospital

Department of Developmental Medicine

Telephone: (03) 9345 5898

Email: dev.med@rch.org.au

Department of Neurosurgery

Telephone: (03) 9345 5437

Email: rch.neurosurgery.org.au

Victorian Paediatric Rehabilitation Service

Telephone: 9345 5283

Email: rehabilitation.service@rch.org.au