What is Pectus Excavatum?

Pectus Excavatum – is Latin and translates as Hollowed Chest and is a chest wall deformity. This is where the breastbone (sternum) is sunken. It is caused by abnormal growth of cartilage (a strong elastic material) between the ribs and the breastbone (sternum). There may be a genetic link to pectus anomalies as it seems to run in families. Pectus Excavatum can appear as a symptom of Marfan syndrome (a genetic disorder of the body’s connective tissue) or sometimes alongside scoliosis (curvature of the spine). Pectus excavatum affects about 1 in 1000 children and is four times as common in boys as in girls.

Pectus Excavatum can be noticeable soon after birth. Although it is often in teenage years that the irregular chest shape is more of a problem as children become more self-conscious. Appearance is the main problem, although in rare cases, it can affect how the heart and lungs work.
What does Pectus Excavatum surgery involve?

The pectus excavatum can be corrected surgically by the Nuss Procedure:

The Nuss procedure was developed by a surgeon in the United States in the 1980’s called Dr Donald Nuss.

Nuss Procedure

This is a newer, less invasive surgical procedure which does not require the chest to be opened. It involves the insertion of a curved steel bar between an incision in either side of the chest and a small incision in the centre of the chest so that the bar lies behind the breastbone. The bar is inserted with the curve facing backwards, and then turned so the curve faces forwards. This pushes out the breastbone so it looks more normal. Sometimes two bars are used. The bar is held firmly in place using a fixation suture around the rib and bar.

This procedure is also known as minimally invasive repair of pectus excavatum (MIRPE).

The Nuss procedure is usually carried out in children and young adults. The operation lasts about 1½ to 2 hours and the bar needs to stay in place for at least 2 years. The bar can be felt beneath the skin, but it is not visible. The incisions are closed with dissolvable stitches.
How well does the Nuss procedure work?

The main outcome measure is satisfaction with the appearance of the chest following surgery. The surgery will improve the shape of your chest which may improve your self confidence and self-esteem.

It is important to remember that it is not possible for us to say to what extent we will be able to improve the shape of your chest with surgery.

Studies looking at the outcome of the Nuss procedure have asked patients to score their appearance before and after the procedure. The UK Register uses a score for appearance on a scale of 1 to 10 (the highest number was 10 denoting a ‘perfect’ appearance).

Data from the UK Register shows an average score of 3.1 before surgery, compared to an average score of 8.4 after surgery.

In a study involving 947 patients, 83% of patients had an excellent result in terms of how they looked. 12% had a good result and 2% had only a fair result. In 2% the pectus excavatum came back, but the bar was removed before 2 years in these cases. There is always a possibility that the anomaly will re-occur once the bar is removed. The level of satisfaction is generally high in other research studies looking at outcomes.

Advantages of the Nuss procedure

The procedure is minimally invasive – only 3 incisions (per bar). So scarring is minimal.

There is no need for cutting or removal of cartilage.

There is generally little blood loss during the procedure. It is rare for a patient having this operation to need a blood transfusion.

Possible risks of the Nuss procedure

NICE has approved the Nuss procedure as a routine treatment option for people with pectus excavatum. As with any surgery, the procedure has risks as well as benefits. Your surgeon will talk to you about these risks, and give you some idea about the chance of them happening:

• Injury to the lungs, heart, liver or an artery in the chest
• Inflammation of the heart tissues
• Fluid collecting in the sac around the heart (pericardial effusion) or around one or both Lungs (pleural effusion).
• The bar moving
• Air or blood in the chest cavity
• Keloid scarring – the scar being red, thickened and itchy
• Wound Infection
• Bone disorders
• **Pain**
• Metal allergy
• Anaesthetic complications

When the risks and benefits of surgery have been explained, you will be asked to go home and think about whether you wish to have the operation or not. The procedure is cosmetic, and does not offer any health benefits, so you need to decide if you think the improvement in appearance is worth the risks involved.
When can the operation be performed?

The surgical correction of pectus excavatum can be performed at any age, but is recommended in mid to late childhood due to growth spurt that happens in late childhood.

We also recommend late childhood as the correct age for pectus surgery so that you are developmentally mature enough to follow the post-op instructions and exercises required for recovery and able to comply with physical activity restrictions, such as restrictions on sports at school etc.

Pre-operative Assessment

Some investigations need to be performed in clinic before listing a child for surgery. These will include a CT Scan, lung function tests, an electrocardiogram and an ultrasound of the heart. The results of these tests are usually normal, unless you have been experiencing symptoms such as shortness of breath on exercise.

What to bring with you

- your admission letter, and any other information we may have sent you
- medicines or inhalers that you are taking at the moment
- toiletries
- nightclothes and a dressing gown
- slippers or comfortable & supportive footwear.
- day clothes - a tracksuit or other comfortable clothes
- glasses or contact lenses (if you have both, please bring glasses as well as contact lenses)

PLEASE SEE THE ALDHERHEY WEBSITE FOR DIRECTIONS TO HOSPITAL AND FOR CAR PARKING TARIFF, THE POST CODE FOR SAT NAV IS L14 5AB.

The day of admission

When you arrive at Alder Hey you should go to the ward 1C. They will prepare you for your operation, and you will be seen by your surgeon and anaesthetist.

The surgeon will explain the surgery again, discuss the risks involved and talk about any worries you may have. If you are under 16 years of age, you will be asked to confirm that you have given your consent for the operation, and your parents will be asked to sign the consent form. If you are over 16 years of age you and your parents can sign the consent form.

The anaesthetist will explain about the anaesthetic and discuss post-operative pain relief with you.

You will be started on a medicine called Gabapentin, which helps with the pain after the operation. You will have routine MRSA swabs done to check for the presence of MRSA bacteria on your skin or in your nose. This test is done for all patients admitted to the hospital and is important in the control of hospital acquired infections.
You will also be seen by a physiotherapist who will explain the exercises you will need following the operation. Compliance with physiotherapy can have a big impact on the rate of recovery following pectus repair surgery, so it is really important that the exercise programme is followed.

**Certain procedures carry an increased risk if the patient is pregnant. All hospitals, including children’s hospitals, are required to check for pregnancy before carrying out procedures such as anaesthesia, x-ray and surgery. This applies to girls aged 12 years or older**

**The Operation**

The operation will be done under general anaesthesia. This means that you will be put to sleep. The surgery will probably last between 1½ to 2 hours.

**What to expect immediately after surgery**

Following your surgical procedure you will be cared for in the recovery department until you are fully awake as you will feel quite drowsy from the anaesthetic. We will monitor your heart rate, blood pressure and oxygen levels and your pain will also be assessed at this time. You will receive a chest x-ray whilst in the recovery department to check the position of the bar and to check there is no air leak (pneumothorax) in either lung. You will then be transferred to 1C to continue your recovery.

**Pain relief following surgery**

To help you feel more comfortable after the operation, nerves may be blocked temporarily by injecting anaesthetic directly onto the nerve roots, near to the spinal cord. This blocks the pain signals and makes them numb. This will be inserted in theatre while you are still asleep.

Strong pain relief medication will also be given straight into the bloodstream. This will be through a cannula, which is a small plastic tube inserted into a vein, usually in the back of your hand. This is called a PCA (Patient Controlled Analgesia). The PCA will allow you to push a button which releases a small dose of pain medication. The machine is programmed to allow a specific dose and the timer on the machine controls how often the medication can be dispensed, even if you push the button more frequently. Oral (by mouth) painkillers will also be used regularly throughout the admission.

*Pectus excavatum surgery is a painful operation and it may not be possible to take away all of the pain in the early days after surgery.*

The medication will help, and things will become more comfortable as recovery progresses. However, you should be aware that you are unlikely to be completely pain free during the first few days after the operation.

On day 2 or 3 post operatively the nerve blocks will be removed by the nursing staff on the ward.

By about the 3rd or 4th day after surgery you should feel quite comfortable on regular oral medication (paracetamol, diclofenac and morphine).

The pain management team will make regular visits to assess your pain and to ensure you have the best pain control medication.
Pain medications may cause side effects such as nausea, vomiting, constipation or sedation/excessive sleepiness. The team looking after your care can order medications to manage these side effects.

**Post-operative recovery**

You will be encouraged to drink as soon as you have recovered from the anaesthetic. If you can take a drink without being sick, then you will be offered something to eat.

Nausea is a possible side effect of anaesthesia, and medication for nausea will be given to help with this, if necessary. You should be coping with a light diet within a couple of days of surgery. Intravenous fluids will be given until you are eating and drinking.

Antibiotics will be given after surgery to help prevent infection. Regular laxatives will also prevent constipation. Constipation is common with intravenous pain medication. This can be very uncomfortable following pectus excavatum surgery, so we ask that parents encourage their children to take their laxatives.

You will be re-positioned regularly during the hours after the operation, and you will be encouraged to move around as soon as you are able. Physiotherapists will make regular visits and give you some information on exercises to help with breathing and circulation, plus upper limb and postural correction exercises will be started the day after your surgery. These exercises will be progressed over the following days and you will be expected to undertake these exercises independently between physiotherapy sessions. You will be encouraged to sit out of bed on the day after surgery and then start walking around as soon as you are able.

**Possible problems & complications**

Minor problems can often occur after operations, including wound infections, chest infections etc. This may be unpleasant, but can easily be treated. Any of these post-operative complications should be completely resolved by 2-3 weeks after the operation.

The more serious complications will have been discussed with you in clinic. These are not common, but if any of these occur, the treatment will be explained to you by your surgeon.

Very occasionally, the metal bar used to correct the sunken chest can move after surgery. This is very unusual, and not dangerous. It can be uncomfortable, but it is a simple procedure to put it back into the correct position.

**Going home**

Patients can usually be discharged 4-5 days after surgery. Physiotherapy will need to be happy with your mobility and upper limb range of movement before going home.

It is very important that you continue with the exercises advised by physiotherapy when you return home.

If you have any questions or queries you can contact the physiotherapy Department on: **0151 252 5862**.

You will be given paracetamol and diclofenac to take home on discharge. Continue both regularly for the first 2-3 weeks after surgery to minimise any pain.

Once discharged, you will be expected to slowly resume normal but restricted activity. Most children are able to return to school within two weeks, with exercise restrictions.
You will need to take it easy for the first four to six weeks after surgery.

- Do not fully stretch both arms together behind your back, at the same time.
- Do not lift or push up from a chair / bed using your arms for 3 months.
- Swimming and jogging can resume after 3 months following surgery.
- No karate, judo, gymnastics, Rugby or other physical / contact sports for the first six months.
- Avoid lifting or pushing of heavy objects for the first three months.
- No backpacks for three months.
- We strongly encourage walking for exercise and should be done frequently to build up your strength. You should begin walking as soon as they get home.
- After the operation and for the first month home, remember to bend at the hip. Do not slouch or slump down when sitting. Good posture will help keep the bar in place.
- You will need to sleep on your back for the first four weeks after surgery, avoid sleeping on your stomach or sides.
- You may bathe or shower once you are out of bed and stable on your feet.
- Wound dressings and (Steri-strips) on the incisions can be completely removed after 5 days from day of discharge.
- CPR can be performed if needed. Compressions will need to be delivered with more force because of the steel bar.
- Defibrillation for cardiac arrest may be performed if needed. Front to back defibrillation pad placement is necessary while the bar is in place.
- No MRIs of the chest and thorax. For MRIs on other areas of the body, you should make the radiologist aware of the pectus bar in order to make a determination of the safety of having the test.
- CT scans are recommended if imaging is needed of the chest.
- Do not drive for three months following surgery. You will need to discuss when you can return to driving with your surgeon and your insurance company.
- You must wear a seatbelt at all times – there is no medical conditions which Justify automatic exemption from the law.

Follow-up Arrangements
You will be given an out-patient appointment for 2-3 weeks after discharge.

Further appointments will be given at 3 months, 6 months and every 12 months until bar removal. A further admission will be required for surgery to remove the metal bar from the chest. This will normally be at least 2 years after the pectus excavatum surgery.

Admission for Pectus Bar Removal – you will be admitted to 1C as a day case surgery.

You will go to theatre usually around 8.30 am. Following the procedure you will go to the recovery department until you are fully awake and then you will go back to the ward to continue your recovery. Later the same day you will be assessed for discharge home.
Important information for Parents

When should I call the Office?

If your child is having any problems and you are worried and would like advice call Mr Dhannapuneni’s secretary

Mrs Emma Burns, Monday - Friday during the day on 0151 282 4514

After hours and on weekends call 0151 228 4811 and ask the hospital operator to bleep (309) for the surgical registrar on call.

IF YOUR CHILD IS SHOWING SIGNS OF :

• Temperature greater than 38°C
• Redness, swelling, drainage or bleeding from the incisions
• Worsening chest pain, especially with deep breaths
• Pain not controlled with pain medications
• Problems with breathing
• Cough that does not go away
• Injury to the chest that may have caused the bar to move
• Sudden onset of chest pain or difficulty breathing

YOU SHOULD ATTEND A & E TO BE ASSESSED

OR CALL 999 AND GET AN AMBULANCE IN AN EMERGENCY.

Our aim is to provide a high quality service to our patients and we welcome any suggestions you may have to help improve our service.
BEFORE SURGERY

AFTER SURGERY

Numbers of Pectus surgical procedures performed by Mr Dhannapuneni
At Alderhey Children’s Hospital

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<tr>
<th></th>
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<th>Ravitch Procedure For Pectus Carinatum</th>
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Less number of pectus operations performed in 2016/17 due to high demand on providing more cardiac operations during that year.
Mr Ram Dhannapuneni

Qualifications
- MBBS: Andhra University, India,
- MBA (Health): Keele University
- FRCS: Royal College of Surgeons in Edinburgh
- FRCS Cardiothoracic: Royal College of Surgeons in Edinburgh.

Memberships
- Society of Cardiothoracic Surgeons in Great Britain and Northern Ireland.
- British Congenital Cardiac Association.
- Chest Wall International Group.
- European Association for Cardiothoracic Surgery.

If you have any Questions or Concerns, please contact:
Mr Ram Dhannapuneni, Consultant Paediatric Cardiothoracic Surgeon:
Tel: 0151 282 4514

This leaflet has been written by Marlene Riley, Surgical Care Practitioner, Cardiac Surgery.

Further information is available from www.pectus.org.

A leaflet is also available from The National Institute for Health and Clinical Excellence (NICE) explaining the guidance it has given on the use of the Nuss procedure for the correction of pectus excavatum surgery. This can be accessed on their website: www.nice.org.uk

This leaflet only gives general information. You must always discuss the individual treatment of your child with the appropriate member of staff. Do not rely on this leaflet alone for information about your child’s treatment. This information can be made available in other languages and formats if requested.

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