Welcome to the first 2019 Physiotherapy Congenital Heart Newsletter. As always, we look forward to hearing from all members regarding fresh ideas and contributions to our newsletter.

**Adult Congenital Heart Disease Module 2019**

Following recent discussions, we agreed there appears to be limited formal training for UK Physiotherapists working with congenital heart patients. I am thrilled to inform PCHN members of an ACHD Module open to nurses and AHPs working in the congenital heart field.

This 2019 course has limited spaces and is very popular nationally. It is also the only accredited module in the UK that focuses on Congenital Heart Disease. This is a unique opportunity and invaluable to support our development and understanding of this highly specialised patient population.

For further details see page 11 of the PCHN Newsletter.

Just a reminder, all CSP and HCP members are entitled to enrol for membership and attend our annual conference. As an added bonus we are free.

For registration please email: Louisa.nielsen@uhs.nhs.uk

**PCHN Twitter account**

http://twitter.com/

Username: PhysioCHN

Please kindly take the time to like our page, post, retweet and comment. This will help our Network to become more established.

**Contributors:** Louisa Nielsen, Caroline Evans and Karen Caulfield

Please do not hesitate to contact Louisa Nielsen if you have any questions or suggestions relating to this newsletter: Louisa.nielsen@uhs.nhs.uk
Case Study: Plastic Bronchitis

Medical and surgical advances for children born with congenital heart defects is evolving. More complex surgeries and improved outcomes mean more patients are surviving into adulthood. What does this mean for us as therapists? A patient’s experiences and journey throughout childhood shape their views and interactions as adults which may impact on their engagement with therapy.

Physiologically the surgical procedures which have enabled patients to live longer also come with side effects and potential complications.

This case study looks at a 13 year old boy with a failing Fontan circulation who was diagnosed with plastic bronchitis and required urgent listing for cardiac transplantation.

Master C was born at term. He was diagnosed at his 20 week scan with double inlet left ventricle (DILV), transposed great arteries (TGA) and a coarctation of aorta (CoA). (please see diagram 1).

![Diagram 1](image)

He was transferred to the cardiac intensive care unit on day one of life and from there underwent the following operations:

<table>
<thead>
<tr>
<th>AGE</th>
<th>Operation / intervention / medical</th>
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<tbody>
<tr>
<td>6 days</td>
<td>CoA repair and pulmonary artery banding via thoracotomy</td>
</tr>
<tr>
<td>3.5 months</td>
<td>Damus procedure (biventricular VSD and aorta repair) Required ECMO support post procedure and an extended PICU stay (3 week)</td>
</tr>
<tr>
<td>10 months</td>
<td>Glenn repair (improve blood flow through the lungs) Extended PICU stay due to pulmonary hypertension</td>
</tr>
<tr>
<td>4 years</td>
<td>Total Cavopulmonary connection (TCPC) / Fontan procedure Chylothorax post surgery requiring 6 month fat free diet</td>
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Master C was seen by the physiotherapist throughout his admissions to ensure that he had opportunities to develop and meet his normal developmental milestones (head control, sitting balance, walking, social interaction etc) as well as to ensure his chest was monitored and managed as required.

His family regularly reported they were finding it difficult to manage Master C’s condition. They had regular psychological support.

Master C had regular clinic appointments and cardiac catheters to monitor his condition often missing school to attend these appointments.
Main problems identified from clinics were:

- Ongoing respiratory infections from the age of 2. On prophylaxis antibiotics over the winter periods effectively managed this
- Ongoing anxiety and behavioural problems in particular difficulty with concentration and anger. (this may be linked to master C’s time on ECMO and his suboptimal circulation due to his cardiac condition
- Mother and grandparents very anxious. Father is not involved in Master C’s life
- Some difficulties at school with tiredness, reduced exercise tolerance and concentration

Master C has been seen in the joint respiratory / cardio clinic regularly and at the age of 12.5 year he presented at clinic with long term temperatures, cough, lethargy. Chest x-ray (CXR) showed right upper lobe changes. At this point he was taught chest clearance techniques (ACBT) and had a home physio programme.

1 week later Master C coughed up a cast and the diagnosis of plastic bronchitis was made. The cast he coughed was approx. 1 cm long, but they can be much bigger as seen in the example in diagram 2.

**What is plastic bronchitis?**

It is diagnosed by expectoration of rubbery casts from the airways. The rubbery casts are caused by lymph fluid leaking into the alveoli from the lymphatic system and building up.

It is rare condition but ominous with poor prognosis and between 1-4% post Fontan surgery is thought to develop the disease due to suboptimal circulation.

**How does it occur / what is lymph?**

Lymph fluid transports fat and protein as well as plays a crucial role in the immune function. It is carried to the veins by the lymph vessels where it is returned to the blood stream.

Plastic bronchitis is caused by a lymphatic flow disorder, an abnormal circulation of lymph fluid or leakage of the fluid which may be due to:

- Injury to thoracic duct (Chris had had a chylothorax post surgery some years ago)
- Congenital abnormalities (Chris has complex cardiac abnormalities)
- Excessive high venous pressures (Chris has high pulmonary pressures) (see diagram 3)
Case Study: Plastic Bronchitis cont...

Master C was known to have pulmonary hypertension and was on regular doses of Sildenafil throughout the day to reduce the pressures in his lungs. Formation and subsequent expectoration of casts is an identification of worsen condition and failing of his Fontan circulation.

Once the cast had been expectorated his chest was clear and his CXR much improved and he did not have on going secretions.

He had an exploratory bronchoscopy 3 months after his first cast which showed a small amount of easily removable casts and minimal secretions. He was started on nebulisers and a home physio regime with an Aerobika device to use 2 x daily (both mum and Chris reported this was not something he did every day). At this point he was assessed for cardiac transplantation given that his circulation was failing, and he was placed on the routine list for transplant.

2.5 months later Master C then presented to A&E with shortness of breath, increased work of breathing and cough. His CXR showed right upper lobe collapse (see diagram 4)

![Diagram 4](image)

Physio input from admission and regularly throughout the days included:
1. Master C’s usual nebuliser and physio regime (Aerobika)
2. Positive pressure with a Clearway
3. Oscillatory pressure with Metaneb

None of these techniques had an impact on his clinical presentation or his CXR over the next few days so he underwent a bronchoscopy and removal of a large cast. Master C was then commenced on Milrinone and admitted to the unit. He was also placed on the urgent list for transplantation.

Post bronchoscopy his chest was clear, he was mobilising well, and his respiratory status was normal.

There is very little in the evidence to suggest to how best to manage patients like Master C. The circulatory flow problem remains until a new heart is found which will improve circulation and then reduce the pressures in the lungs (this can take a number of months to normalise). This means that a cast will reform but the timeframe of this reformation is unclear. Between cast formation his lung fields are clear on auscultation and CXR.

As a team we discussed the best management to ensure his lungs are in as optimal health as possible.
Case Study: Plastic Bronchitis cont...

Problems identified whilst on the unit awaiting heart transplantation:
- Poor compliance with nebuliser and physio regime
- Anxiety and difficult behaviours
- Parental anxiety
- Awaiting cardiac transplantation as an in patient on HDU as attached to an intravenous delivery of inotrope. This meant that he could not see his friends daily or attend his usual school and activities. His home was not close to the unit. Chris felt like he had no freedom. The timeframe for this is unknown, a transplant could be available in days, months, hours. It is totally unpredictable.

Clinical presentation:
- Independently mobile on the ward and able to go to the parent accommodation within the hospital
- Nebuliser plate changes to ensure the nebuliser delivery was as effective and fast as possible
- Jointly negotiated a routine for Master C to use his Aerobika straight after his nebulisers.
- Chest clear on auscultation, CXR clear
- Strong effective dry cough

Thorough assessment is needed should any respiratory parameters change (increased cough, reduced saturations etc.) and appropriate chest clearance techniques can be used. Up to the point of change physiotherapy intervention has been on a weekly monitoring basis and Master C continues to have a nebuliser and Aerobika regime to follow.

The MDT working with Master C aim to give him a feeling of control back in some aspects of his life to improve compliance and behaviour.

Key learning points:
- What plastic bronchitis is
- How congenital cardiac condition may give rise to it
- The complexity of holistic management for children born with a congenital cardiac condition
- Consideration as to how living with a long-term condition may affect development, relationships and effect compliance

If you have any questions or would like to find out any further information, please contact me on the details below.

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Atrial Septal Defects

Atrial Septal Defects (ASD’s) are a direct communication between the cavities of the atrial chambers, which permit shunting.

They are one of the most common congenital heart disease defects as an associated lesion occurring in about 6 to 10% of all cardiac malformations.

ASD’s are more common in females (2:1).

There is a well recognised association of Atrial Septal Defect (ASD) with Down Syndrome (secundum or primum).

**Secundum ASD** - defects of the oval fossa (most common 60%).

**Superior sinus venous ASD** - deficiency of the atrial wall at the superior vena cava (SVC), the SVC connects to both atria. It is not a true defect of the atrial septum (15%).

**Inferior sinus venous ASD** - override the Inferior vena cava. This form of ASD is less common.

**Coronary sinus ASD** is a communication between the left atria and the coronary sinus which is the rarest type.

**Primum ASD’s** is the atrial component of an AVSD (20%).

The size of the ASD and the compliance of the right ventricle and pulmonary vascular bed determine the degree of intra-atrial shunting.
Associated lesions
When ASD is the primary diagnosis, associated malformations occur in 30% of cases and include:

- Partial anomalous pulmonary venous connection
- Pulmonary valve stenosis
- Mitral stenosis or mitral valve prolapse
- Ventricular septal defect
- Patent ductus arteriosus
- Coarctation of the aorta

Presentation and course in childhood
Most children with an ASD present with a murmur and are asymptomatic.

Occasionally, infants may present with breathlessness, recurrent chest infections and even heart failure.

Children with sizable ASD’s and right heart dilatation should undergo elective closure of their defect for prognostic reasons during the first decade of life, irrespective of symptoms.

Course in Adulthood
Most adults present with symptoms usually in the third or fourth decade of life. Usually breathless on exertion and/ palpitations due to atrial tachyarrhythmias. This often correlates with an increase in left to right shunting.

Occasionally, adults may present with cardiac enlargement on routine chest x-ray or a heart murmur. The latter type of presentation is particularly common among pregnant woman reflecting increased circulating plasma volume.

Adults with ASD’s have reduced survival if ASD closure takes place after 25 years of age. Other late complications of unrepaired ASD’s are right heart failure, recurrent pneumonia and pulmonary hypertension, atrial flutter and fibrillation and paradoxical embolus and stroke.

Management options for adults with ASD
Management is determined by the size and type of defect, indications for closure of ASD are as follows:

ASD with cardiac enlargement on x-ray, a dilated right ventricle on echo and a pulmonary artery systolic or mean pressure 50% or less than the corresponding aortic pressure. Patients should be considered for elective closure irrespective of age or symptoms.

Younger and older patients benefit from ASD closure compared to medical therapy in terms of:

- Survival
- Functional class
- Exercise tolerance
- Reduction of risk of heart failure
- Reduction of risk of pulmonary hypertension
Atrial Septal Defects cont...

However, patients older than 40 years of age and particularly those with preoperative rhythm disturbance remain at risk of sustained atrial arrhythmia after closure. Surgical or transcatheter ablations should be considered.

History of TIA or stroke in presence of ASD or persistent foramen ovale and right to left shunting.

Contraindications to closures include pulmonary vascular resistance of more than 7-8 units or a defect diameter of less than 8mm with no evidence of right heart dilatation and asymptomatic.

All secundum defects should be considered for transcatheter closure. Defects up to 40mm can be closed by an occulder! Very large oval fossa defects can be closed only by surgery using cardiopulmonary bypass with potential for greater morbidity in the elderly with arrhythmias.

Device closure outcomes
Intermediate results are comparable to surgery with few major complications.

Functional capacity improves and supraventricular arrhythmias are better tolerated with more response to medical management.

Occasionally residual atrial septal defects are encountered either after catheter or surgical closure. Unless responsible for a significant left to right shunt, generally they do not require additional intervention.

Longer follow up is needed to determine the incidence of arrhythmias and thromboembolic complications later after device closure.

Surgical outcomes
Secundum ASD’s without pulmonary hypertension should undergo closure with a very low (<1%) operative mortality.

Early and long term follow up is excellent.

Pre-existing atrial flutter and fibrillation may persist unless arrhythmia targeting procedures are performed.

Medical management
Management of associated complications include: right heart failure, atrial tachyarrhythmia and occasionally pulmonary hypertension.

Pregnancy and contraception
Pregnancy is usually well tolerated by most women with an un-operated atrial septal defect. Cardiology review is recommended because of small risk of paradoxical embolus and stroke, arrhythmia and heart failure. If circumstance allow, ASD’s should be closed prior to pregnancy. Only contraindication to pregnancy in women with ASD’s is persisting pulmonary hypertension.
Late complications

- Premature death
- Right heart failure
- Left ventricular dysfunction
- Tricuspid and mitral valve regurgitation
- Atrial flutter/fibrillation
- Sinus node dysfunction
- Paradoxical thromboembolism
- Endocarditis (rare)
- Systemic arterial hypertension
- Pulmonary hypertension/pulmonary vascular disease (usually a very late complication)
Resources

AHA/ACC ACHD guidelines 2018 https://www.acc.org/guidelines

American Heart Association https://www.heart.org/

Adult Congenital Heart Association https://www.achaheart.org/

ACHD Learning Centre.org http://achdlearningcenter.org/category/learning-modules/
ACHD Learning Centre http://achdlearningcenter.org

British Heart Foundation www.bhf.org.uk

British Journal of Cardiac Nursing www.magonlinelibrary.com/toc/bjca/current

Canadian adult congenital heart network http://www.cachnet.org/index.cgi

Cincinnati Children’s hospital https://www.cincinnatichildrens.org/patients/child/encyclopedia/defects

Cove Point Foundation http://www.pted.org/?id=home


*European Society of Cardiology www.escardio.org


Leeds ACHD Centre www.essentialachd.org

National Institute for Health and Care Excellence www.nice.org.uk

Resuscitation Council UK www.resus.org.uk

Somerville Foundation www.thesf.org.uk

The CHiP Network www.chip-network.org

The International Society for Adult Congenital Heart Disease (ISACHD) www.isachd.org
Courses

Postop morphology of the congenital heart 2019
Date: 25th to 26th February 2019
This series of workshops is aimed at providing doctors and allied healthcare professionals with an understanding of the morphological changes in the hearts of patients who had previous surgical/interventional procedures for congenital heart defects.
Venue: Royal Brompton Hospital
Contact: morphology@rbht.nhs.uk

Wessex Congenital Cardiac Series Study Day 2019
Date: March 22nd, 2019
Topics will be AVSD and Ebstein’s: Everything you need to know! Live demonstration of morphology specimens
Venue: University Hospital Southampton
Contact: abigail.sharpe@uhs.nhs.uk

Hands-on Cardiac Morphology Courses 2019
3 day course suitable for all those wishing to improve their knowledge of structural heart disease
Date: 10th to 12th April 2019
Venue: Royal Brompton Hospital
Contact: morphology@rbht.nhs.uk

17th Introduction Congenital Heart Disease 2019
Date: 8th to 12th July
Venue: Evelina Children’s Hospital, St Thomas
Contact: CHDweek@gstt.nhs.uk

Adult Congenital Heart Disease Module 2019
A 20 credit module at Level 6 or 7
Date: Variable (commencing 25th April 2019)
Venue: Southwark, London
Contact: https://www.applycpd.com/LSBU/courses/1_103578?tabId=1 (level 6)
https://www.applycpd.com/LSBU/courses/1_103558?tabId=1 (level 7)

ISACHD - Core Curriculum Course in Adult Congenital Heart Disease
To facilitate achieving core curriculum requirements, trainees (and consultants) can access an online course devised by the International Society for Adult Congenital Heart Disease (ISACHD).

Please do not hesitate in contacting Louisa Nielsen if you have any questions or suggestions

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