PedsCases Podcast Scripts

This is a text version of a podcast from Pedscases.com on "**Congenital CHF.**" These podcasts are designed to give medical students an overview of key topics in pediatrics. The audio versions are accessible on iTunes or at www.pedcases.com/podcasts.

Congenital Congestive Heart Failure

Developed by Jyote Boora and Dr. Angela McBrien for PedsCases.com.
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Introduction:

My name is Jyote Boora and I am a third-year medical student from the Faculty of Medicine and Dentistry at the University of Alberta. This podcast was supervised by Dr. Angela McBrien, a pediatric cardiologist at the Stollery Children's Hospital in Edmonton, Alberta. This podcast will broadly cover congenital-related congestive heart failure in children. A future podcast will cover non-congenital related etiologies of congestive heart failure in children.

The objectives of this podcast are as follows:

1. Define Congestive Heart Failure (or CHF), and describe its epidemiology and classification.
2. Discuss the pathophysiology of CHF in children.
3. Describe the typical signs and symptoms of CHF by different age groups, as well as typical findings on physical exam.
4. Demonstrate the utility of the various diagnostic tests for CHF.
5. Outline the various treatments for CHF.

To understand what CHF looks like in clinical practice, let's begin with a case to illustrate the initial presentation of this condition.

A 2-month-old infant named Max is brought in to see his regular pediatrician for his two-month checkup. His parents mention that over the past month, they have noticed that their son has had decreased oral intake that is progressively becoming worse. They first noticed this 1 week after he was born.

It took some time for his parents to notice his poor oral intake as he had other issues occurring at the same time. In the first few days of life he experienced jaundice for which he was treated with phototherapy. Other issues that arose in the first 3 weeks of

his life included a tongue tie release and circumcision. His parents tried increasing the frequency of feeds, from every 6 hours to every 4 hours. In addition, every time he feeds, they note that he sweats profusely, as if it's a hot summer day. Although he seems to be outwardly happy and smiling, his parents notice that he is breathing quickly at times and has abdominal and intercostal indrawing, which seems to be getting worse over time. Additionally, he often vomits after he feeds, approximately 2-3 times per day. This has been exhausting for his family as they also have an older 7-year-old daughter who goes to school and is involved in numerous activities.

Max's mother is a G3P2, who has a history of gestational diabetes treated with insulin during her pregnancy. She had adequate prenatal care during pregnancy, and was compliant in taking folate and multivitamins. All prenatal screening tests, including a routine prenatal ultrasound were negative. Max was delivered at 37 weeks via elective c-section. His APGAR scores were 8, 9 and 9. His mother tried to breastfeed, but to no avail as he struggled with latching on to the breast. As a result, his parents have been feeding him via bottle with either formula or expressed breast-milk. Max just had his first immunization, on time at two months of age. He has a healthy older sister who is 7 years old. Of note, there is no family history of congenital or acquired heart disease.

We will return to this case later on, to review the physical exam, diagnosis and management of CHF.

Epidemiology and Classification

CHF occurs when the heart is unable to pump enough blood to meet the body's oxygen and metabolic demands. CHF is estimated to affect between twelve thousand and thirty-five thousand children below the age of 19 each year in the United States. There are different classification systems used to stratify heart failure. One such system typically used with adults is the New York Heart Association or NYHA scale. This scale assigns a score from 1 to 4 based on the patient's symptoms. A score of 1 is for a patient with no limitation of physical activity, fatigue, or dyspnea, whereas a score of 4 would mean that a patient is unable to carry out any physical activity without experiencing discomfort, and has symptoms of heart failure at rest.

Another system used particularly for infants and children is the Ross classification system. The Ross classification system was developed to provide a more global assessment of heart failure severity in all pediatric age groups. It incorporates feeding difficulties, failure to thrive and symptoms of exercise intolerance that somewhat resembles the NYHA scale.

The Ross scale goes as follows:

Class 1: The patient is asymptomatic.

Class 2: The patient has symptoms of mild tachypnea. If they are an infant, they experience diaphoresis with feeding. If they are an older child, they are experiencing dyspnea on exertion.
Class 3: The patient has symptoms of marked tachypnea. If they are an infant, they have marked diaphoresis with feeding. If they are an older child, they are experiencing marked dyspnea on exertion. In infants this also includes prolonged feeding times with growth failure.
Class 4: The patient has symptoms such as tachypnea, intercostal or abdominal indrawing, grunting or diaphoresis at rest.

Pathophysiology

CHF is caused by either a problem of too much volume in the heart (or preload), too much pressure in the heart (or afterload) or ventricular dysfunction.

Excess volume results in an inability for the ventricles to eject all the blood they receive, as there is simply too much fluid for the heart to effectively pump out with each contraction. Congenital-related causes of volume overload include a valvular defect, such as mitral regurgitation or a left-to-right shunt such as a Ventricular Septal Defect (VSD), or Patent Ductus Arteriosus (PDA).

Excess pressure (or afterload) can result in impaired ventricular contractility, and congenital-related causes of this include aortic stenosis, or coarctation of the aorta.

Ventricular dysfunction is where the ejection of blood from the ventricle to the systemic vasculature is impaired. This can occur in children with structurally normal hearts or those with congenital heart disease. As this podcast focuses on congenital related heart disease, we will focus on those etiologies as opposed to structurally normal hearts.

However, causes of CHF in structurally normal hearts include: cardiomyopathy, myocarditis, myocardial infarction, infection, arrhythmias, drugs, and other non-cardiac causes such as chronic kidney disease.

In congenital-related CHF, as we have mentioned, ventricular dysfunction is often secondary to increased afterload (such as that caused by a semilunar valve obstruction or coarctation).

However, it can occur in relatively rare structural conditions, such as the condition Anomalous origin of the Left Coronary Artery from the Pulmonary Artery (ALCAPA), which results in myocardial ischemia or infarction of the left ventricle.

Signs and symptoms

The signs and symptoms of CHF in children vary based on the age of presentation. For example, going back to our case with Max, he had symptoms of diaphoresis with feeding, poor oral intake, vomiting, and tachypnea.

More common symptoms in infants can include: Poor oral intake due to breathlessness, reflux or vomiting, failure to thrive or poor weight gain, tachypnea, or diaphoresis with feeding. If the CHF is related to a left-to-right shunt, the symptoms are often absent in the newborn period and gradually appear and worsen over the first weeks and months of life.

Infants are unlikely to develop dependent edema, as they spend a lot of their time recumbent. Tachycardia is a common sign of CHF in infants, and they may have palpitations, but of course they cannot complain of their palpitations and so typically tachycardia is noted on physical exam as opposed to heart palpitations. Cyanosis is not associated with CHF in infants, unless they are in extremis or have right-to-left shunting as part of their condition.

Symptoms in older children and adolescents more commonly include fatigue, exercise intolerance, dyspnea, orthopnea, abdominal pains, nausea and vomiting. Less common are symptoms of chest pain, palpitations, edema or ascites.

As you can see, recognition of heart failure in children without any prior heart disease can be challenging, because these symptoms are quite nonspecific, and more often in children these symptoms can have non-cardiac causes. Therefore, it is important to have a high index of suspicion for CHF when you see children presenting with these nonspecific symptoms.

Of course, family history is important - however, most infants with CHF do not have any prior family history, so the absence of this should not stop you from including CHF in your differential diagnosis.

Back to the two month checkup with our patient, Max. On physical exam, you notice that he is demonstrating poor weight gain and growth velocity on his growth chart. Using your stethoscope, you auscultate over Max's left lower sternal border to hear a Grade 1/6 holosystolic murmur.

On physical exam of a patient with suspected CHF, here are some key things to look for:

For inspection and vitals: The triad of tachycardia, tachypnea and hepatomegaly are classic for CHF. In addition to tachypnea, look for accessory muscle use and abdominal and intercostal indrawing as markers of respiratory distress. Additional clues to respiratory distress in infants are grunting or nasal flaring as they breathe. Do not forget

to feel the femoral pulses, which are reduced, absent or delayed in coarctation of the aorta. In older children, they may present with wheezing.

Please note: that poor perfusion, as seen with cool mottled extremities, reduced capillary refill, reduced peripheral pulses and hypotension are signs of end stage CHF.

For auscultation: an S3 gallop may be heard due to volume overload. Listen for a murmur, while not always present this can be heard in most cases with structural congenital disease that results in CHF. The quality and location of the murmur can give you clues about the underlying structural congenital defect which is causing CHF. For example: a holosystolic murmur at the lower left sternal edge usually indicates a VSD (or more rarely, tricuspid regurgitation). Don't forget, that large VSDs cause soft murmurs, so a soft murmur can be more worrisome than a loud one. In our case, recall that you noted a soft, grade 1 out of 6 murmur.

Furthermore, systemic congestion findings may include: hepatomegaly, ascites, and peripheral edema. Again, remember that in infants, findings of peripheral edema and ascites are rare.

Diagnostic Tests

After you perform your physical exam on Max, you have a strong suspicion about what is going on with him. To confirm your suspicions, you request a Chest X-Ray as well as an Electrocardiogram or ECG. The X-ray shows no pulmonary effusions, however there is mild cardiomegaly and left ventricular hypertrophy, which is corroborated the ECG results.

Tests that are useful for diagnosing congenital related causes of congestive heart failure includes the following:

Chest X-ray is indicated as a first line investigation in children with suspected heart failure. The absence of cardiomegaly reduces the likelihood of heart failure and has high sensitivity.

ECG is also part of the first line series of investigations for CHF. It can point towards the cause of CHF such as biventricular enlargement in the case of VSDs, left ventricular hypertrophy in the case of aortic stenosis, or an atrial, junctional or ventricular tachycardia as either an underlying cause or complication of heart failure.

Echocardiography is used to assess heart chamber size, myocardial function, ejection fraction, and to diagnose structural congenital heart defects that cause CHF. ECG and echo are typically the first line tests ordered by pediatric cardiologists, with less reliance upon Chest X Rays.

As it turns out, Max is referred to pediatric cardiology and seen 2 months later. An echocardiogram shows a 9mm apical muscular VSD, with mild mitral regurgitation (caused by the volume load on his left heart) and a mildly dilated left atrium.

Management and Treatment

Once a patient is diagnosed with a congenital heart defect complicated by CHF, first-line treatment is directed at optimizing preload and afterload accordingly. In the case of our patient, Max, the cause of his CHF turns out to be a VSD as confirmed by the echocardiography. One of the main principles of management is to increase calorie intake, as infants with CHF will have higher calorie needs due to higher metabolic demands. Therefore, a pediatric dietician consult is important. Ideally, we want to optimize Max's weight gain which will make Max a better surgical candidate and improve his post-operative outcome.

In order to reduce preload, diuretics such as Furosemide may be used. Furosemide reduces preload by dumping sodium into the urine via the ascending loop of Henle. Spironolactone may also be given to compensate for the loss of potassium via loop diuretics, in addition to its own diuretic properties.

Although this is not a consideration for our patient Max, inotropes may also be used, such as digitalis, dopamine, and dobutamine for patients in whom there is impaired cardiac contractility impacting their CHF, such as in cases of aortic stenosis, or coarctation of the aorta.

Agents to reduce afterload include ACE inhibitors such as captopril, which is commonly used for treatment of a VSD. Milrinone may also be used as an IV infusion, typically in more acute care settings such as Pediatric ICU.

Returning to our case...unfortunately, after being seen by pediatric cardiology ,Max's condition worsens over the next two weeks, as the natural history of the disease progresses. The patient's poor oral intake has worsened, and the vomiting has increased. The sweating has increased to the point where he is drenched in sweat, soaking his clothes and the sheets in his crib. Max's parents bring him back to the emergency department, where he is subsequently admitted for closer monitoring and management. A naso-gastric(or NG) tube is placed to facilitate feeding and optimal weight gain. His Furosemide dose is doubled, which helps control his symptoms for the most part. He is also started on captopril, which greatly reduces his symptoms.

From here, management will depend on the cause of the congenital-related CHF. For example, with Max's case, management will depend on the size and location of the VSD. For moderate size VSD's, it may be reasonable to observe and wait for spontaneous closure. Otherwise, open surgical repair or percutaneous repair, or some combination of the two may be considered.

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Most large VSDs are managed with a primary surgical repair. In Max's case, the VSD was in the apical part of the septum, and would have represented a challenging case for surgical closure. Therefore, the options in his case would be as follows:

Option 1: Undergo placement of a pulmonary artery band in order to reduce his symptoms of heart failure by restricting pulmonary blood flow. Then, wait for his VSD to reduce in size over time, and have a repeat operation to remove the band, or

Option 2: Undergo a hybrid procedure – which is a combination of interventional and surgical approaches to try to close the VSD with a device such as an Amplatzer.

If a large left-to-right shunt is left unrepaired long term, then pulmonary hypertension will develop, due to chronic excessive pulmonary blood flow caused by the right-to-left shunt. In the worst case scenario, patients can develop Eisenmenger syndrome, which consists of irreversible pulmonary hypertension causing a right-to-left shunt. For large VSDs, it is recommended to undergo closure before 6 months of age in order to avoid this complication.

Ultimately for Max, it is decided that the hybrid approach would be the optimal treatment and this took place when he was 5 months old. The procedure involved a midline sternotomy and placement of an Amplatzer device to occlude the VSD. The procedure lasts about 3-4 hours long. Post-operatively, he does well besides a short course of fever which is corrected with antipyretics. After 4 days in hospital, he is discharged.

Max is recovering well and is completely free of his previous symptoms. Upon follow-up 1-week post-op, an echocardiogram shows a 2-3mm residual VSD. However, his mitral regurgitation has completely resolved. He is still having trouble with oral feeding, and a pediatric dietician is consulted. He is started on Aspirin 20mg daily to prevent any clots forming around the device and has another checkup in 9 months' time to monitor his remaining VSD, which is projected to close on its own as he grows.

Conclusion

Let's summarize the key points from this podcast:

- Congestive Heart Failure (or CHF) is a progressive condition occurring when the heart is unable to keep up with either increasing volume overload, pressure overload, or ventricular dysfunction.
- Congenital causes of CHF in children include Ventricular Septal Defect, Patent Ductus Arteriosus, and Coarctation of the Aorta.
- Symptoms vary across patient age groups. In infants, common signs and symptoms include poor feeding with vomiting/reflux, hepatomegaly, tachypnea, diaphoresis with feeding, and failure to thrive. In older children, patients can

present with exercise intolerance, orthopnea, nausea and vomiting, peripheral edema and abdominal pain.

- There are three useful diagnostic tests for CHF and its congenital-related causes: 1. Chest X-Ray, 2. Electrocardiogram, and 3. an Echocardiogram.
- Treatment for congenital-related CHF focuses on using pharmacologic agents to reduce afterload and preload, such as Furosemide and Captopril as well as optimizing growth. Once the congenital cause of the CHF is discerned, appropriate surgical or percutaneous management is pursued should the patient be eligible.
- Finally - it is important to maintain a high index of suspicion when seeing children presenting with the aforementioned signs and symptoms as the presenting features of congenital-related CHF are nonspecific and can be easily missed if you are not considering it in your differential diagnosis.

Thank you for listening to this Pedcases podcast series on the topic of congenital-related congestive heart failure in children.

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