

Hypoplastic Left Heart Syndrome: An Unexpected Complication

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Objectives

- Highlight the disease pathology of hypoplastic left heart syndrome.
- Showcase the importance of maintenance care by highlighting a complication of the disease.

Introduction

- Hypoplastic left heart syndrome(HLHS) is a congenital heart disease that involves malformation of the left side of the heart.
- HLHS effects 1 in every 4,344 babies born in the United States.
- The disease can be diagnosed on prenatal ultrasound or shortly after birth.
- No cure for the disease has been identified and the goal of treatment is to decrease the burden of the disease.
- Intervention must be provided early to increase survivability.
- Complex cases of HLHS often require heart transplant.
- A variety of complications have been associated with the disease.

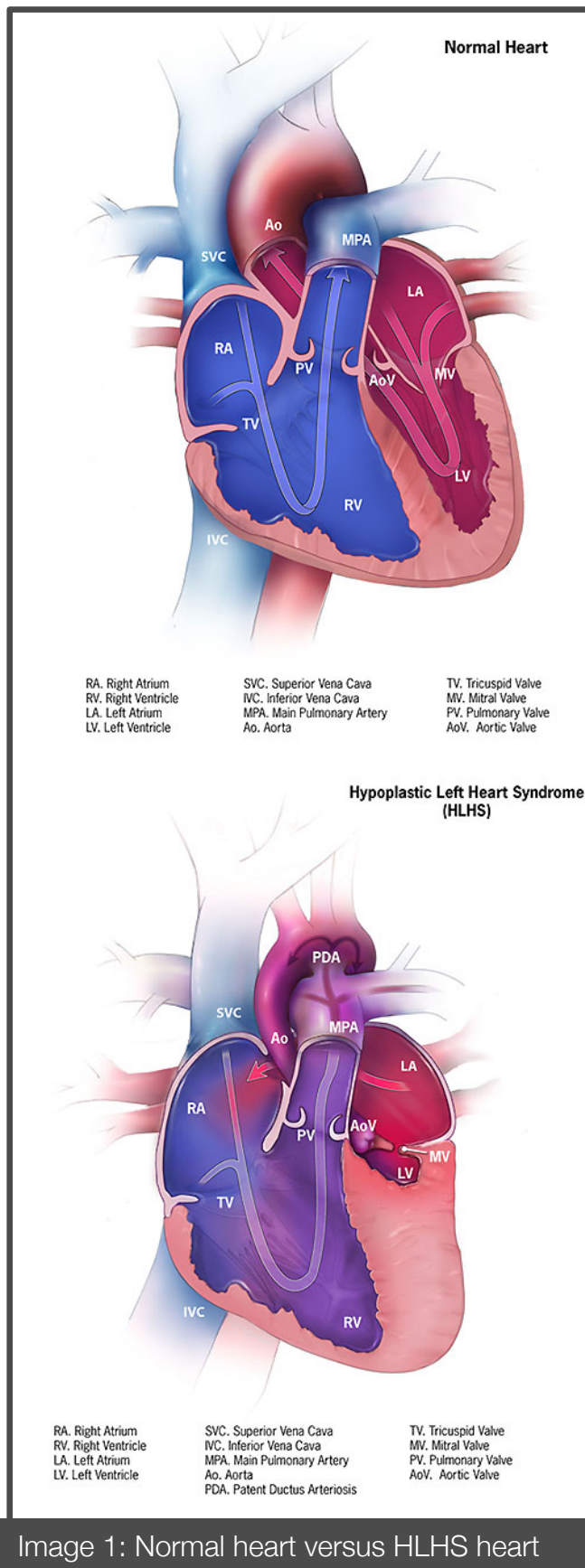


Image 1: Normal heart versus HLHS heart.

Medical History

- 4 year old male patient born with hypoplastic left heart syndrome.
- Fontan procedure and epicardial dual chamber pacemaker placed within first month of life.
- Patient is not a candidate for transplant due to multiple comorbidities.
- Comorbid conditions include:
 - NYHA Class II systolic congestive heart failure
 - Pulmonary hypertension
 - Gastrostomy tube
 - Chronic vomiting and diarrhea
 - Dysphagia
 - GERD
 - Cerebral infarction, thrombosis of cerebral artery
 - Autonomic neuropathy

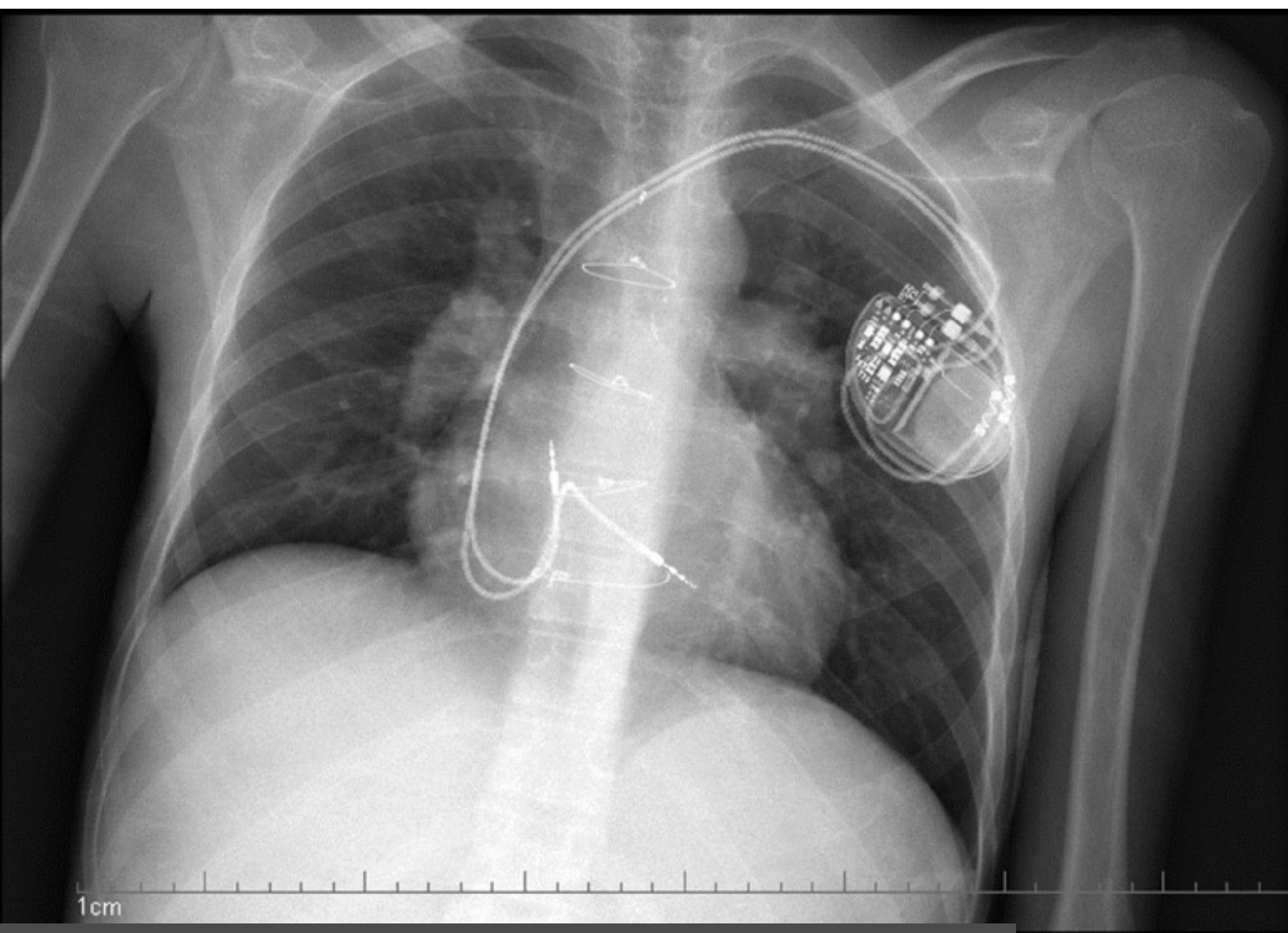


Image 2: Radiography of epicardial dual chamber pacemaker in pediatric patient

Case Study

INITIAL ENCOUNTER

History:

- 4 year old male presents to the pediatric clinic with painless abdominal swelling onset 1 day ago.
- Denies fever, chills, chest pain, abdominal pain, nausea or vomiting.
- CBC, CRP and ESR done 1 day prior, all results within normal limits.

Physical Exam:

Vitals: BP: 94/60 HR: 114 RR: 26 Temp: 97.7 F O2: 89%

General: Awake, alert and in no acute distress. Not acutely ill appearing.

Abdomen: Abdominal wall swelling noted in the RLQ medial to the pacemaker. No pain elicited on palpation.

Diagnostics:

Abdominal Ultrasound:

- RUQ shows 2 small fluid collections near pacemaker leads measuring 5 and 6 mm.
- RLQ shows focal bulge measuring 3 cm.

Management:

- Obtain ultrasound
- Consult Cardiology
- Watchful waiting, return to the clinic if symptoms worsen



SUBSEQUENT ENCOUNTER

History

- Patient returned 7 days later with persistent abdominal swelling now complicated by pain and skin color changes onset 1 day ago.
- Patient's mother states that he is denying pain but his expressions display pain.
- Patient's mother unable to get into contact with cardiologist.
- Denies any fever, chills, chest pain, abdominal pain, nausea or vomiting.

Physical Exam:

Vitals: BP: 102/64 HR: 116 RR: 24 Temp: 96.4 F O2: 88%

General: Awake, alert and in no acute distress. Not acutely ill.

Abdomen: Abdominal swelling consistent with the formation of an abscess.

Patient does not complain of pain on palpation.

Diagnostics:

Wound cultures: Growth of Staphylococcus epidermidis

- Susceptible to: Clindamycin, Daptomycin, Trimethoprim/Sulfamethoxazole, Vancomycin

CT Chest:

- Fluid collection adjacent to pacemaker wires in anterior upper abdomen measuring 16 x 12 x 14 mm.
- Fluid near but not obviously surrounding pacemaker leads.
- Ultrasound recommended for further evaluation.

Ultrasound Abdomen:

- Complex fluid in pacemaker pocket measuring 4.6 x 4.3 x 0.5 cm with some mobile debris. Nearly subcutaneous fluid collection.
- Mild edema/inflammation in surrounding soft tissues.

Blood cultures: Negative for growth of any organisms

Management:

- Patient admitted to inpatient care
- IV Vancomycin
- Pacemaker removal with external cardiac pacing
- Routine labs drawn, all cultures negative 3 weeks after pacemaker removal.
- Pacemaker replaced. Labs, cultures and radiography negative for infection.
- Discharged home on Daptomycin to complete 6 week antibiotic regimen.

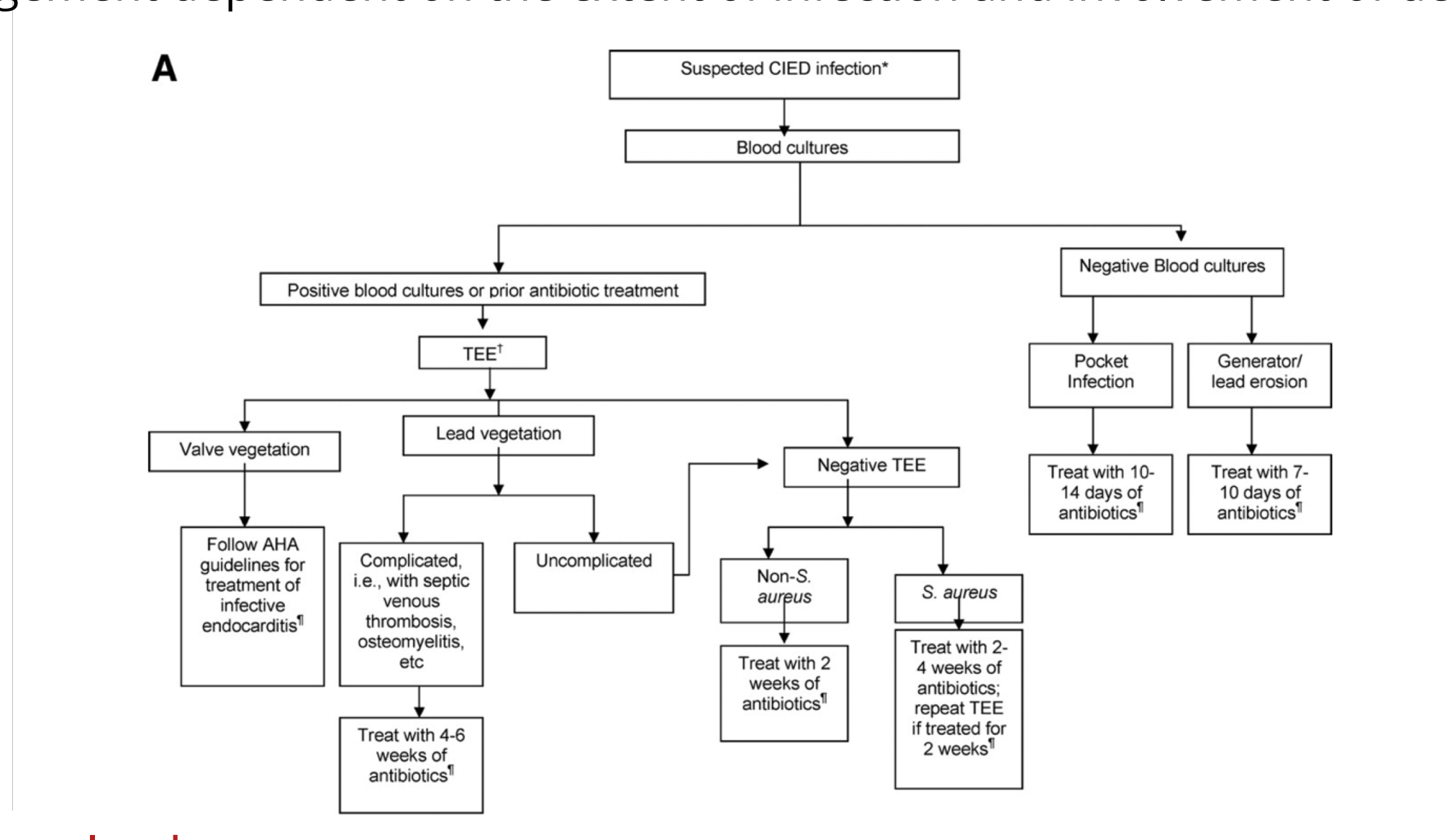


Discussion

- Patient had multiple risk factors for infection including heart failure, compromised immune system and history of repeated cardiac catheterization.
- Pediatric patients with pacemakers due to congenital heart conditions have a 1% - 8% rate of infection, 30% of which is from epicardial pacemakers.
- Patient not volunteering pain could have caused the infection to progress further.
- Early diagnosis and management majorly abated the development of sepsis.
- Barriers to treatment that were overcome in this case:
 - Communication between rural pediatrician and children's hospital to assure proper treatment in a timely manner.
 - Access to walk-in clinic for monitoring of disease progression.
 - Electronic medical record imaging, allowing for better documentation of disease.

Ideal Management

- Empiric antibiotic treatment with IV Vancomycin.
- Most common etiology is Staphylococcal infections.
- Obtain 2 sets of blood cultures at initial evaluation
- Transthoracic echocardiogram to assess for any signs of infectious endocarditis
- Management dependent on the extent of infection and involvement of device



Conclusion

- Cardiac pacemaker infections do not always have an obvious presentation.
- The pediatric population is at an increased risk of infection, necessitating close monitoring.
- Prompt and aggressive treatment of infection is imperative to improve patient outcomes.
- Communication between primary care and specialty providers ensures the proper management of disease processes.

References

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Center for Disease Control and Prevention. (n.d.). Congenital Heart Defects - Facts about Hypoplastic Left Heart Syndrome | CDC. Retrieved from <https://www.cdc.gov/ncbddd/heartdefects/hlhs.html>

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- Image 1: <https://www.cdc.gov/ncbddd/heartdefects/hlhs.html>
- Image 2: <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3634247/>
- Diagram A: <https://www.ahajournals.org/doi/10.1161/CIRCULATIONAHA.109.192665>

