

## AN HIV-POSITIVE MOTHER'S CHILD EXPERIENCING A CYANOSIS CRISIS: AN ECONOMIC BURDEN

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### Abstract

A 6-year-old healthy male subject is brought to the emergency department ICU. His parents report that this child has been suffering with these symptoms since he was 10 months old. Though the subject was born to an HIV positive mother he was HIV negative. He has a cough and difficulty breathing. Upon arrival, the child appears to be ill and in significant respiratory distress. Subject suffering from Dextrocardia, large posterior upper muscular VSD, MLV to MRV shunt (difficult routability to AORTA), aortic from RV, No antegrade pulmonary flow, confluent branch PA's, smooth phasic unobstructed flow in the BD Glenn shunt, good LV function, and IVC uninterrupted. Dextrocardia is a condition in which the heart is pointed toward the right side of the chest. Normally, the heart points towards the left. The condition is present at birth (congenital). The parents report that their child has been coughing for a few days without significant sputum production. He has also seemed slightly more fussy than usual and has had a poor appetite and mild nasal congestion over the past few days. He and his parents are taken immediately to a resuscitation room, and the resident physician and attending physician are called to the bedside by the ED staff. The child has no known allergies. Although his appetite is poor, he was tolerating feedings up until today. The parents do not report any known sick contacts or travel. Depending on the cause, treatment could involve medications, oxygen therapy, surgeries, or specialist consultations. These can all be financially draining. Parents used to miss their work to care for their child's medical needs, leading to lost wages. Frequent doctor visits and hospital trips can add up in transportation costs. Depending on the

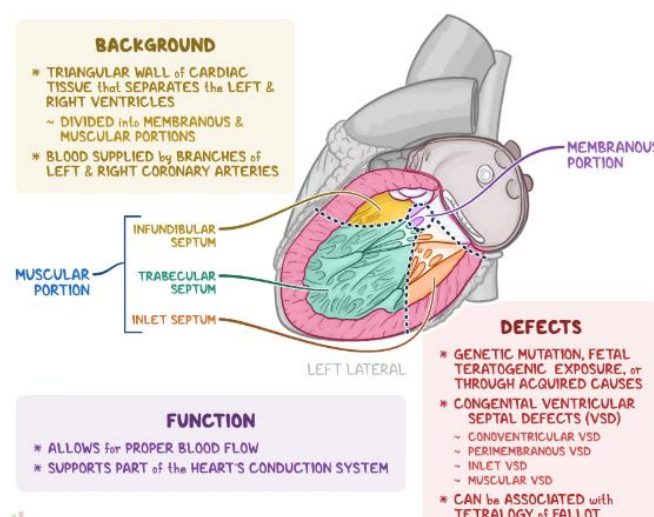
cause of cyanosis, the child might require special equipment like nebulizers, oxygen concentrators, or wheelchairs, adding to the financial strain. The stress of caring for a sick child can take a toll on parents' mental health, potentially impacting their ability to work or function normally.

**Keywords:** Dextrocardia, large posterior upper muscular VSD, MLV, MRV shunt, loss of wage, financial drain.

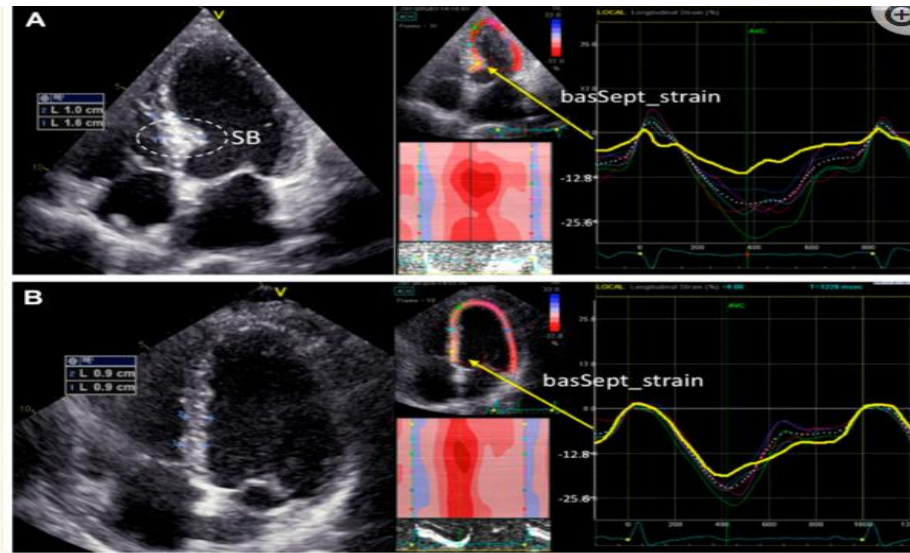
## Introduction

Ventricular septal defect (VSD) is the most common congenital cardiac anomaly in children and is the second most common congenital abnormality in adults, second only to a bicuspid aortic valve. An abnormal communication between the right and left ventricles and shunt formation is the main mechanism of hemodynamic compromise in VSD. While many VSDs close spontaneously, if they do not, large defects can lead to detrimental complications such as pulmonary arterial hypertension (PAH), ventricular dysfunction, and an increased risk of arrhythmias.<sup>[1,2,3]</sup> VSDs were first identified by Dalrymple in the year 1847.<sup>[4]</sup>

Ventricular septal defects (VSDs) are the most common congenital cardiac anomaly in children and the second most common congenital abnormality in adults, second only to bicuspid aortic valves. The main mechanism of hemodynamic compromise in ventricular septal defects occurs due to abnormal communication between the right and left ventricles and shunt formation. This activity reviews the presentation and pathophysiology of ventricular septal defects and highlights the role of the interprofessional team in the management of patients with congenital heart defects.



Source: <https://www.osmosis.org/answers/interventricular-septum>



Source:

Figure 2

Examples of segmental longitudinal systolic strain curves derived from speckle tracing imaging in patients with (A) or without (B) septal bulge (SB). Gaudron PD, et al. J Am Soc <https://www.ncbi.nlm.nih.gov/pmc/articles/PMC9181036/>

The interventricular septum is an asymmetric curved structure due to the pressure difference in ventricular chambers. It is composed of five parts: the membranous, muscular (frequently referred to as trabecular), infundibular, atrioventricular, and the inlet. <sup>[4,5]</sup>

### Those who suffered an episode of endocarditis.

When the ratio of the pulmonary blood flow to the systemic blood flow ( $Q_p/Q_s$ ) is equal to or more than 2 plus clinical evidence of LV fluid overload.

In milder shunts such as those with  $Q_p/Q_s$  above 1.5, it is reasonable to intervene when there is evidence of LV systolic or diastolic dysfunction, or when the pulmonary artery pressure and pulmonary vascular resistance are less than two-thirds of systemic pressure and systemic vascular resistance, respectively.

Surgical repair reduces the risk for endocarditis, might improve PAH, and overall it increases survival. Without PAH, the operative mortality rate is approximately 1%. Complications include residual or recurrent VSD, valvular incompetence such as tricuspid

regurgitation and aortic insufficiency, arrhythmias, LV dysfunction, and progression of PAH. The arrhythmias which accompany VSD repair include atrial fibrillation, complete heart block, and uncommonly, ventricular tachycardia. The main contraindication for surgical VSD closure is the presence of irreversible PAH; this is due to the high surgical perioperative mortality and pulmonary complications.

Percutaneous device VSD closure is reserved for those whose surgery is very risky due to severe PAH, multiple comorbidities, and those who had prior cardiothoracic surgery such as residual or recurrent VSD. Muscular VSDs are the main type amenable to this procedure, the proximity of other defects to the inlet valves makes performing this technique challenging in such cases.

In conclusion, VSD is the most common congenital anomaly at birth. Small defects are expected to close spontaneously in the first year of life; however, larger defects can result in severe complications. Surgical VSD closure and device closure are the main intervention for large defects.

### **Presentation of Case**

This is a retrospective study. The Subject was born to HIV positive parents who are taking ART and during the time of pregnancy she used Acyclovir for the treatment of Herpes infection.

We examined child and family characteristics, identified through literature review and a priori theory, as potential confounders of the association between CHD and financial burdens and mental health needs, as well as potential predictors among CSHCN with CHD. Child characteristics included age, sex, race/ethnicity, insurance type (any private, public, uninsured), medical home status, effect of condition on daily activities compared with children of the same age, and type of comorbidities (physical-only, developmental-only, physical and developmental, or none). Comorbidities were defined as a parent reporting ever being told by a provider that their child had at least one of the following conditions: physical conditions (asthma, diabetes, epilepsy or seizure disorder, migraines or frequent headaches, blood problems, cystic fibrosis, cerebral palsy, muscular dystrophy, arthritis or joint problems, allergies, and head injury, concussion or traumatic brain injury), or developmental conditions (attention deficit disorder or attention deficit hyperactivity disorder, depression, anxiety,

behavioural or conduct problems, autism or an autism spectrum disorder, developmental delay, an intellectual disability or mental retardation.

He is suffering from Dextrocardia, large posterior upper muscular VSD, MLV to MRV shunt (difficult routability to AORTA), aortic from RV, No antegrade pulmonary flow, confluent branch PA'S, smooth phasic unobstructed flow in BD glenn shunt, good LV function, IVC uninterrupted.

Dextrocardia is a condition in which the heart is pointed toward the right side of the chest. Normally, the heart points toward the left. The condition is present at birth (congenital).

Weight: 12.1kg      Temperature: 98F      Heart Rate: 120/bpm  
Respiratory Rate: 24/min      SpO2: 73%      Head Circumference: 47cm  
Fall Score: (9)

**BRIEF COMPLAINTS & HISTORY OF PRESENT ILLNESS**

- K/C/O CHD: Diagnosed as CHD in neonatal period, complex congenital heart disease. Underwent Glenn shunt with MPA ligation and atrial septectomy in Hyd on 7/3/18
- Cyanosis: Increases while crying
- DOE: Class II
- Family H/O: nothing significant
- Medication history: Ecosprin 75mg

**GENERAL EXAMINATION**

- Clubbing grade 2
- Cyanosis present

**SYSTEMIC EXAMINATION**

- Cardiovascular  
Remarks: apical impulse in left 5th ICS in MCL, S1 S2: ESM grade 2 at LLSB. No gallop.
- Respiratory  
Remarks: chest clear

**DIAGNOSIS**

- Corrected transposition of great vessels (disorder), Primary, Provisional, 19/10/2020
- Remarks: S/P BD Glenn at Hyderabad on 7/March/2018

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**AGNOSIS**

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**EDUCATION ORDER**

DRUG NAME	DOSE	DOSEAGE
1) ASPHRIN TABLET 75MG: ECOSPRIN	D - 1 - 0	Start Date: Sep 06, 2021, Once Daily, (10:00-12:00) Tablet, After Food for TELL REVIEW Instruction - Do not stop the medication without consulting your doctor
2) IRON-HYD B12-FOLIC ACID SYRUP 100MG: TCMOFERON	3ml 1 - 0 - 0	Start Date: Sep 06, 2021, Once Daily, (13:00-01:00) ml, 1/2 Hour Before Food for TELL REVIEW Instruction - Do not stop this medicine without consulting physician

**TESTS**

- INVESTIGATION: Echo: Pls see report. S, L, L, DEXTROCARDIA, LARGE POSTERIOR UPPER MUSCULAR VSD, MLV TO MRV SHUNT (DIFFICULT ROUTABILITY TO AORTA), AORTIC FROM RV, NO ANTEGRADE PULMONARY FLOW, CONFLUENT BRANCH PA'S, SMOOTH PHASIC UNOBSTRUCTED FLOW IN BD GLENN SHUNT, GOOD LV FUNCTION, IVC UNINTERRUPTED
- ECG: Sinus Tachycardia, Superior Axis, RV Forces
- CXR: Normal CXR, Low Op

could → Pg 3

**Fig.1: Clinical summary of the subject.**

## Discussion

The Case Challenge series includes difficult-to-diagnose conditions, some of which are not frequently encountered by most clinicians but are nonetheless important to accurately recognize.

In the account of the follow-up of Lillehei's pioneering open heart repairs [5], it is reported that complete heart block occurred in 4 of the 27 patients in whom closure was attempted. In



reports collected from the last 30 years, describing series of from 23 to 265 patients, although some had no incidence of complete heart block [6,7,8,9,10], this complication occurred in as many as 4% of the others, with an incidence of as high as 8% reported in one series spanning a period of 21 years.<sup>[11]</sup>

Some children with perimembranous VSD may develop aortic valve prolapse and require surgery. Finally, all unrepaired VSDs have the potential to increase pulmonary vascular resistance leading to Eisenmenger syndrome. At this stage, except for a heart and lung transplant, there is no other viable therapy. With a marked shortage in organs for transplantation, the majority of these patients succumb to progressive right heart failure and cyanosis.

A growing body of literature is addressing the financial burdens and mental health needs of families of children with CHD.<sup>[12]</sup> A few studies have reported general financial difficulties related to having a child with CHD<sup>[13]</sup> as well as specific burdens such as large out-of-pocket expenses and decreases in salary due to reducing hours or stopping work to care for the child.<sup>[14]</sup> Close to 50% of families of children with special healthcare needs (CSHCN) experience some type of financial burden,<sup>[15]</sup> but the magnitude of financial impact of CHD on families is not well documented. Increased risk of caregiver psychological distress, particularly around critical periods of illness such as diagnosis and surgery, persisting up to three years after the newborn period has been documented.<sup>[16,17,18,19]</sup>

## Conclusion

In conclusion, VSD is the most common congenital anomaly at birth. Small defects are expected to close spontaneously in the first year of life; however, larger defects can result in severe complications. Surgical VSD closure and device closure are the main intervention for large defects. Moreover, our study has demonstrated the benefits of transcatheter closure in terms of lower complication rates and mean hospital stay. However, surgery still has a place for the closure of large and complex defects.

Therefore, defect closure should not be postponed to a later age in patients who have symptoms that can be attributed to a left-to-right shunt, and transcatheter closure seems to be a safe and effective alternative treatment for sASD in this population.

## Acknowledgements

## Authors' Contributions

Conceptualization,N.B.J., data curation,N.S.V, and writing—review and editing T.K.

Consent: Written consent has been taken from subject parents.

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