

## Hypoplastic Left Heart Syndrome (HLHS)

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**Section:** Cardiovascular

**Area of Interest:** Arteries / Aorta Cardiac Cardiovascular system

**Procedure:** Surgery

**Imaging Technique:** CT

**Imaging Technique:** CT-Angiography

**Special Focus:** Congenital Case Type: Clinical Cases

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**Patient:** 6 days, male

### Clinical History:

6-day old male neonate presented with cyanosis within 24 hr of birth, respiratory distress, irritability, poor feeding. Examination revealed tachypnea, tachycardia, crepts in bilateral lungs, abnormal cardiac auscultation.-

### Imaging Findings:

Hypoplastic aortic root, ascending aorta (AA), and arch of aorta (AOA) with normal-sized Descending aorta (Fig1). Hypoplastic left ventricle (LV) and ventricular septal defect (VSD) (Fig 2) seen. Normal left arterio- ventricular communication (Fig 3). Long tubular Patent ductus arteriosus (PDA) between AOA, distal to left subclavian artery origin and Left Pulmonary Artery (LPA) (Fig 4). Dilated main pulmonary artery (MPA) is seen towards left side of AA and shows absence of normal crossover (Fig 5). Dilated MPA, Right Pulmonary Artery (RPA) and LPA are seen (pulmonary arterial hypertension changes) (Fig 6). Dilated Right atrium(RA) with Atrial septal defect (ASD) seen (Fig 7). Dilated and hypertrophied right ventricle(RV) with normal right arterio- ventricular communication (Fig 8). All the pulmonary veins draining into left atrium(LA). Superior vena cava (SVC) and inferior vena cava (IVC) draining into RA. IVC and hepatic veins are dilated.

### Discussion:

HLHS is a complex congenital cardiac malformation which if left untreated is, invariably fatal. [1,2]. It is characterized by hypoplasia/aplasia of left-sided heart structures with normal right-sided heart structures and normal ventriculo-arterial concordance. The exact aetiology is not fully known, however, it is believed to be caused by multiple errors during the early stages of cardiogenesis.

During intrauterine fetal development, hypoplastic left-sided cardiac structures i.e. Mitral valve (MV), LV, and aortic valve (AV) cause hypoplasia of aortic root and AA leading to reduced blood flow into AA during systole. However, the presence of PDA allows retrograde perfusion of coronary arteries and upper body region during diastole. Thus in intrauterine life, the fetus thrives well.

However, during extrauterine life, LA is not able to pump adequate blood through stenotic MV, which leads to pulmonary venous outflow obstruction and a typical feature of neonate with congestive cardiac failure. ASD allows left to right shunting of blood which may be restrictive or non-restrictive depending upon the size of ASD. A restrictive pattern of ASD is more common than no restrictive pattern. AV stenosis coupled with LV hypoplasia leads to reduced flow into AA during systole. The right-sided heart then receives as well as supplies all the blood. However, it cannot provide an adequate amount of oxygenated blood (neither functionally nor physiologically) for adequate perfusion, resulting in systemic and coronary ischemia, which causes cyanosis, acidosis, and death if left

Echocardiography is the investigation of choice antenatally (most reliably ~22 weeks) as well as postnatally. It is the most common congenital cardiac malformation detected in fetal life. [4]

The incidence is between 1.7-6.7: 10, 000 live births [5]. CT scan allows direct visualization of vessel anatomy and malformation [6] and provides valuable information for pre-operative planning, however, is less commonly performed due to its inability to assess function, valves, and radiation safety issues.

Treatment is mainly surgical and includes Norwood procedure [7] bidirectional cavopulmonary anastomosis or Hemi-Fontan procedure.

### **Take-Home Message**

Nowadays CT pulmonary angiography is being done for pre-operative evaluation of congenital cardiac anomalies. HLHS is seen to be associated with other cardiac anomalies. Therefore, its recognition is imperative for the radiologist and cardiac surgeon for proper treatment planning and hence surgical correction is equally important for the overall success of surgery.

Written informed patient consent for publication has been obtained.

**Differential Diagnosis List:** Hypoplastic left heart syndrome, Infantile aortic coarctation , Interrupted aortic arch

**Final Diagnosis:** Hypoplastic left heart syndrome

### **References:**

- Norwood wi. Hypoplastic left heart syndrome. Cardiol clin 1989; 7:377-385 PMID: 2659180.
- Norwood wi. Hypoplastic left heart syndrome. Ann thorac surg 1991; 52:688-695. PMID: 1898174
- Bardo dm, frankel dg, applegate ke et-al. Hypoplastic left heart syndrome. Radiographics. 21 (3): 705-17 pmid: 11353117
- Dr. Gurivi reddy.c, shrinuvasan.s, bruntha. D, chidambaram.r1; (2016) imaging of hypoplastic left heart syndrome –a rare antenatal cardiac anomaly. Indian journal of basic and applied medical research vol.-6, issue- 1, p. 20-23.
- Jose m sierra1, md, sandra r silva, md, philippe jeanty, md, ph.d. (2002) hypoplastic left heart syndrome. Sonoworld 07-10-12
- Gilkeson rc, Ciancibello l, Zahka k. Pictorial essay. Multidetector CT evaluation of congenital heart disease in pediatric and adult patients. AJR am j roentgenol. 2003;180 (4): 973-80. AJR am j roentgenol (full text) PMID: 12646439
- Alsoufi b, bennetts j, verma s et-al. New developments in the treatment of hypoplastic left heart syndrome. Pediatrics. 2007;119 (1): 109-17. Doi:10.1542/peds.2006-1592 - PMID: 17200277

**Figure 1**

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**Description:** Sagittal MPR MIP image- showing the course of the entire thoracic aorta. AA and aortic root are hypoplastic with a small-sized AOA and a normal-sized descending thoracic aorta. Note the normal branching pattern from AOA **Origin:** © Department of Radiology, Khetan Diagnostic Centre, Jaipur, India, 2021

**Figure 2**

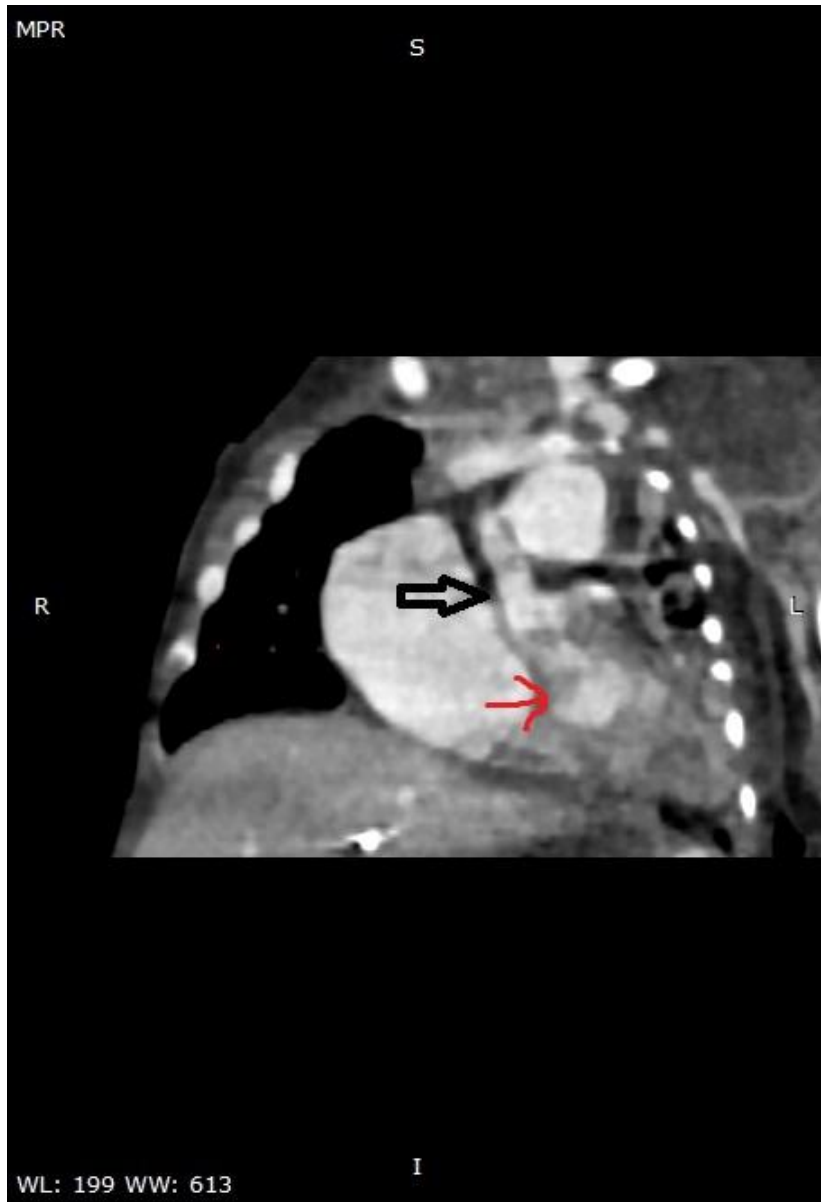


**Description:** Axial CT scan -at Inter-ventricular septal level – showing severely dilated RV with small-sized hypoplastic left ventricle (marked with black arrow).VSD (marked with green arrow). is also seen

**Origin:** © Department of Radiology, Khetan Diagnostic Centre, Jaipur, India, 2021

**Figure 3**

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**Description:** Coronal CT image - showing the origin of the aorta (marked with black arrow) from the hypoplastic left ventricle (marked with red arrow). Note the normal left ventriculo-arterial concordance

**Origin:** © Department of Radiology, Khetan Diagnostic Centre, Jaipur, India, 2021

**Figure 4**



**Description:** Axial CT scan - at arch of aorta level showing large PDA between the right-sided aorta and left-sided dilated pulmonary trunk **Origin:** © Department of Radiology, Khetan Diagnostic Centre, Jaipur, India, 2021

**Figure 5**



**Description:** Axial CT scan- proximal to pulmonary artery bifurcation- showing hypoplastic ascending aorta (marked with arrow). Dilated MPA is seen towards the left side of AA and shows the absence of normal crossover **Origin:** © Department of Radiology, Khetan Diagnostic Centre, Jaipur, India, 2021

**Figure 6**

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**Description:** Axial CT scan-at pulmonary bifurcation -showing dilated MPA, right and left pulmonary arteries. Pulmonary edema and consolidation are seen in bilateral lung fields, predominantly in dependent regions **Origin:** © Department of Radiology, Khetan Diagnostic Centre, Jaipur, India, 2021



**Figure 7**

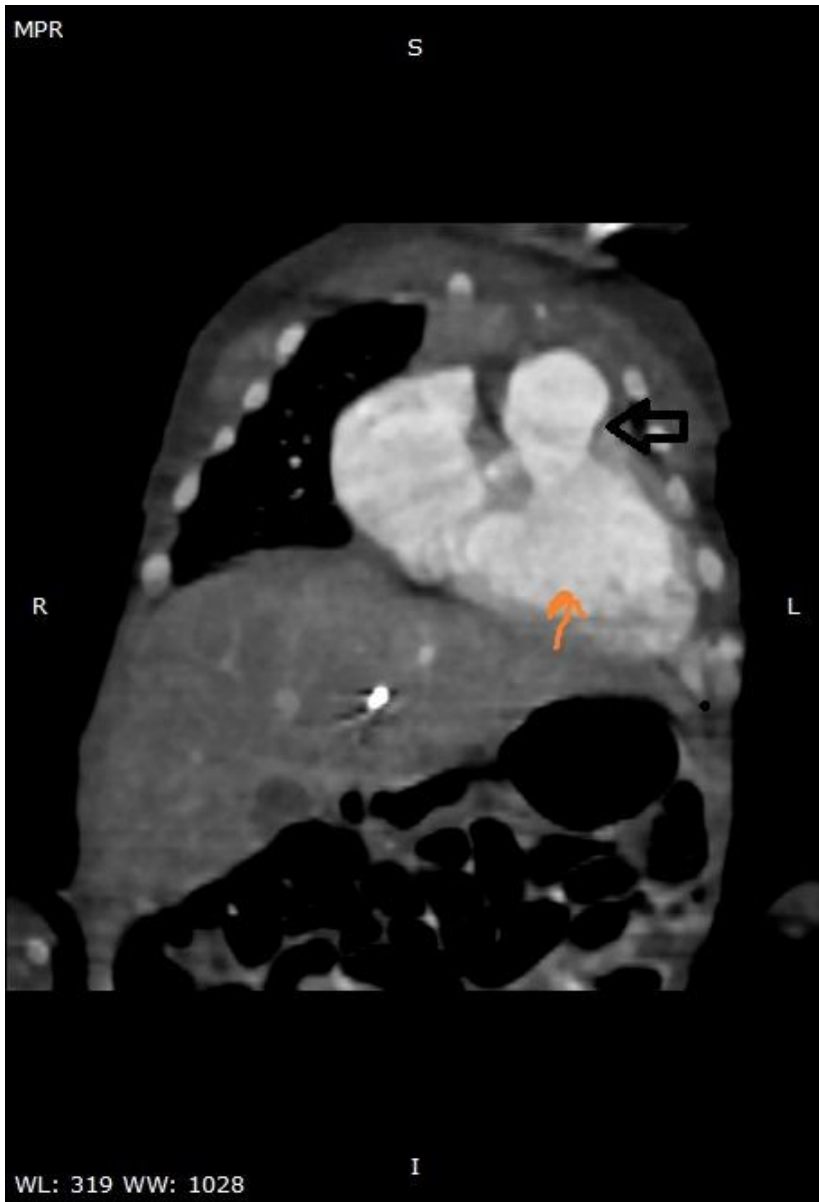
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**Description:** Axial CT scan showing dilated RA (marked with black arrow) and ASD (marked with red arrow) **Origin:** © Department of Radiology, Khetan Diagnostic Centre, Jaipur, India, 2021

**Figure 8**

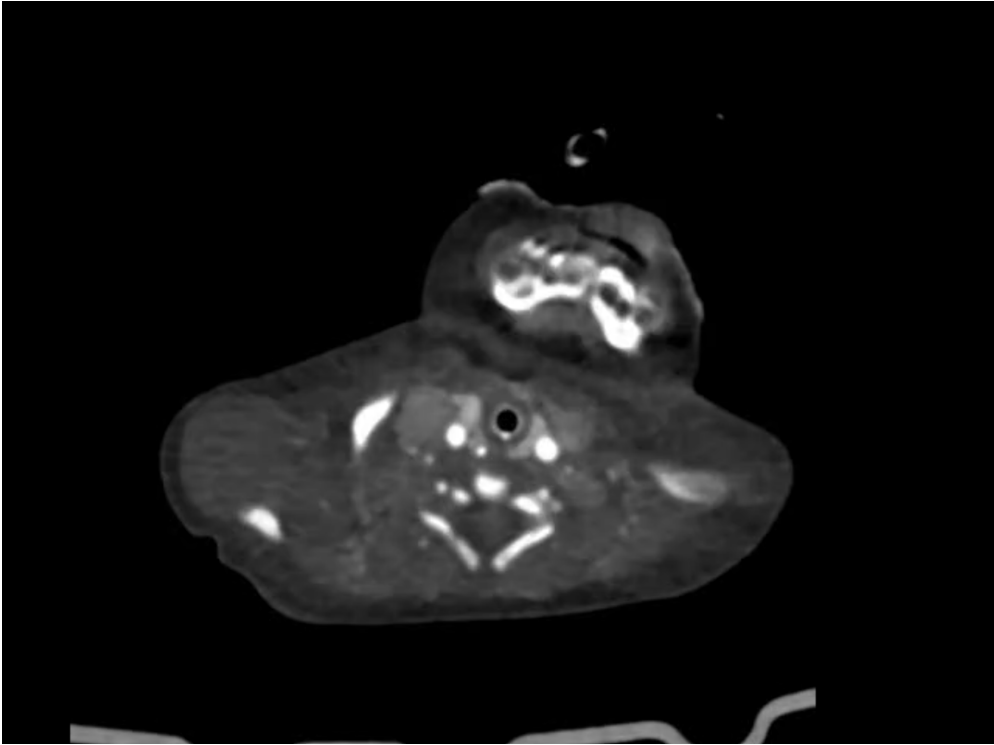
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**Description:** Coronal CT scan- showing normal right ventriculo-arterial concordance with dilated MPA (marked with black arrow) originating from dilated RV (marked with orange arrow) **Origin:** © Department of Radiology, Khetan Diagnostic Centre, Jaipur, India, 2021

**Figure 9**

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**Description:** Axial CT scan showing above findings **Origin:** © Department of Radiology, Khetan Diagnostic Centre, Jaipur, India, 2021