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Case Report

Hypoplastic Left Heart: An Exceptional Singular Case

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Abstract: Hypoplastic left heart (HLHS) includes hypoplastic left heart, aortic atresia with hypoplastic but perforate mitral valve .It accounts for 0.04 to 0.16 per 1000 live birth and there is male prevalence of 55-70 % .It has been called most malignant form of congenital heart disease with an average life span of 5-14 days, with 95% of afflicted infants die within first month of life.
Keywords: Hypoplastic left ventricle, Hypoplastic LV, Congenital hypoplasia of left side.

INTRODUCTION

Congenital Hypoplasticleft heart syndrome(HLHS) include hypoplasticleft heart, aortic atresia with a hypoplastic but perforate mitral valve. It occurs in 0.04 to 0.16 per 1000 live births and accounts for7.5% of infants with congenital heart disease and is responsible for 25% of cardiac deaths in the first week of life .The malformation has been called the most malignant form of congenital heart disease, a conclusion underscored by an average life span of only 5-14days. Precarious survival depends on three variables :patency of ductusarteriouses , pulmonary vascular resistance, and an adequate interatrial communication [1].

Ninety five percentof afflicted infants die within first month of life. There is male prevalence of 55% to 70% in aortic atresia with a hypoplastic but perforate mitral valve. Maternal age tends to be above average with amean of 31 years. A mosaic chromosomal 22q11 deletion is associated with HLHS and genetic disorders include Turner syndrome, Trisomy 13, Trisomy 18 and trisomy 21[2].

CASE REPORT

In this exceptional case, a baby boy of 4 month age born through normal delivery presented with history of respiratory distress of 6 day duration along with failure to thrive and poor feeding history, bluish discoloration of lips, fingers and tongue and mild degree of fever since the past 6 days. The patient had history of repeated upper respiratory infection during the past 4 months for which she consulted doctors.

On examination; respiratory rate was 44/minute, heart rate was 150/minute central cyanosis was present, blood pressure was 80/60mmHg, his oxygen saturation was 77%.

Arterial pulsation: His carotid, brachial pulsations were palpable, with striking right ventricular impulse and palpable pulmonary closure sound.

On auscultation he had soft mid systolic murmur.Second heart sound was loud and single.There was presence of third heart S3 sound. His ECG had presence of tall peaked P wave and QRS wave showed evidence of right ventricular hypertrophy. X-Ray shows evidence of cardiac enlargement of Right Atrium and Right ventricle type. There is presence of prominent pulmonary artery and pulmonary congestion.Apart from this he had bilateral non homogenous opacity suggestive of Pneumonia. Echocardiography showed Hypoplastic Left ventricle with atretic aortic valve and hypoplastic Mitral valve. Right ventricle was hypertrophied with presence of large outlet ventricular septal defect with left to right shunt, ascending aorta was hypoplastic and tubular.

Management

The baby was started on inotropes, Fluids and put on assisted ventilation along with Antibiotics. As the baby needed multiple staged surgery he was referred to higher Cardiac centre but unfortunately his hemodynamic instability and respiratory infection compounded his problem and he could not undergo surgery.

Following surgical procedures are considered in patients with Hypoplastic left ventricle:

First stage “Norwood Procedure” establishes communication between Right ventricle and aorta by connecting transacted proximal pulmonary artery and ascending aorta usually with a patch extending around the augmented ascending aorta. Pulmonary artery circulation is established by communication between Right Ventricle and Pulmonary Artery conduit. Patent Ductus Arteriousesis ligated and inter-atrial communication is created.

Second stage consists of Superior vena cava to Pulmonary artery connection (bidirectional Glens

procedure) or a Hemi-fontan. Incorporating the roof of atrium to the pulmonary artery anastomosis, this is performed at 6 month of age as an intermediate procedure before stage three, that is Fontan operation.

The survivor of norwood operation are entering adult life presently. Other option is cardiac transplant.

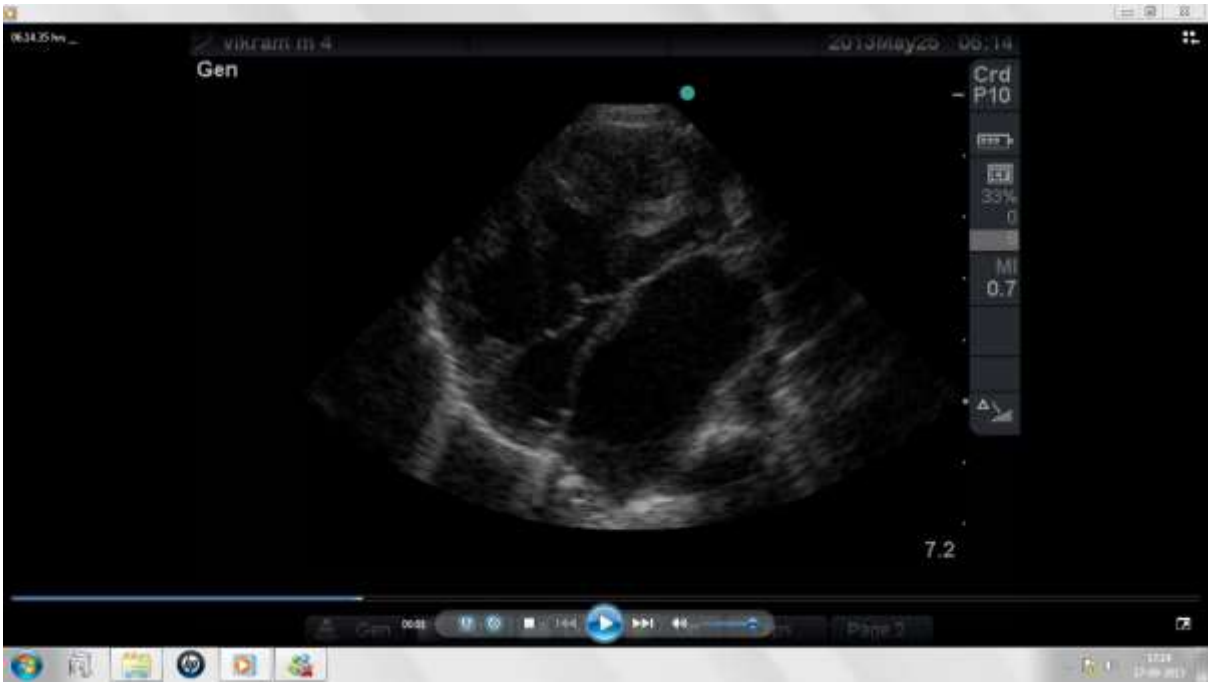

Fig. 1: Picture of Hypoplastic Left Ventricle in A-P 4 Chamber View Withenlarged Right Ventricle

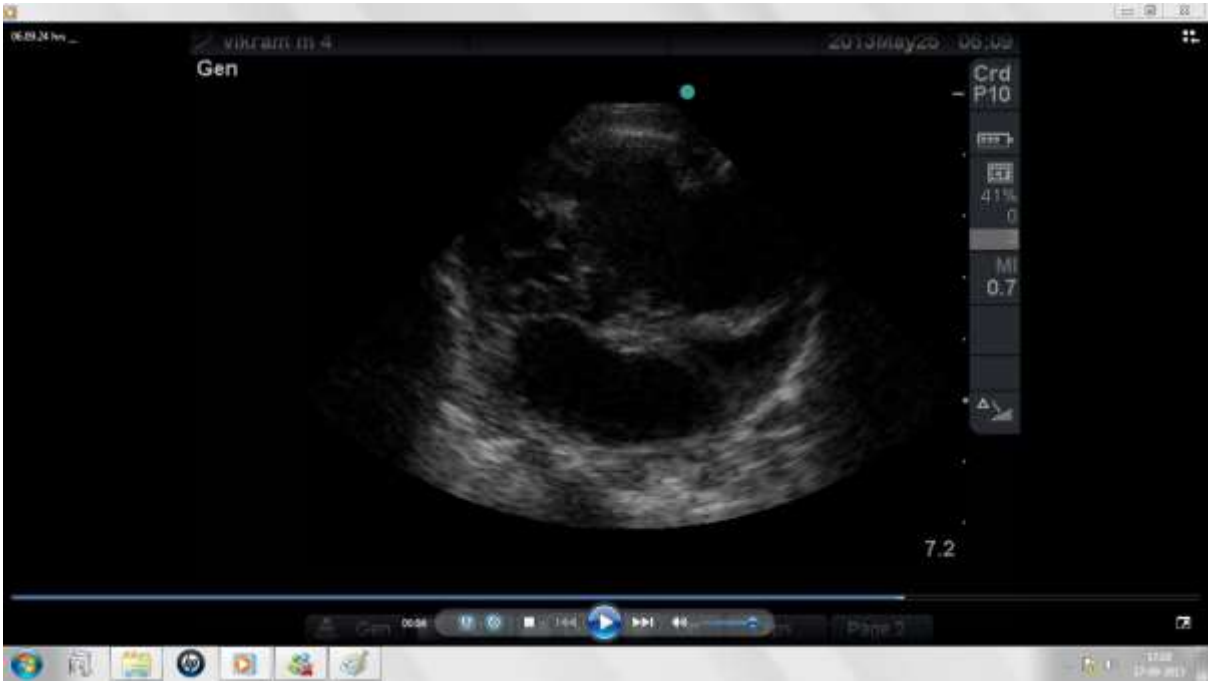

Fig. 2: Picture of Hypoplastic Left Ventricle in Plax View With Large Outlet Ventriculer Septal Defect

CONCLUSION

A patient with HLHS represented by Aortic atresia, a hypoplastic but perforate mitralvalve present as tachypneic, listless and often moribound male neonate with cyanosis that varies from mild to profound.Right ventricular impulse are conspicuous.Auscultation occasionally detects a tricuspid regurgitation murmur, a pulmonary middiastolic murmur,or more often than not no murmur at all. The electrocardiogram displays right

atrial ‘P’ waves and right ventricular hypertrophy. The X ray discloses pulmonary venous congestion and dilation of right atrium, right ventricle and pulmonary trunk but conspicuous absence of ascending aorta. A ventricular septal defect is associated with well-formed left ventricle. The physiologic consequences are the same and longitivity patterns are similar. Echocardiography shows hypoplastic left ventricle lined with endocardial fibroelastoses hypoplastic perforate mitral valve ahypoplastic tubular ascending aorta guarded by an atretic aortic valve .

REFERENCES

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