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A rare case of hypoplastic left heart syndrome in pregnancy and its outcome

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Abstract
Hypoplastic left heart syndrome (HLHS) is an etiologically multifactorial congenital heart disease affecting in 1 in 5000 newborns. This syndrome has been reported to occur in approximately 0.016% to 0.036% of all live births [1, 2].
A Case of primigravida with 35week period of gestation with Gestational hypertension with left hypoplastic heart syndrome with single umbilical artery which was diagnosed at 24weeks anomaly scan but she continued the pregnancy. Emergency ceasarean section was done i/v/o impending eclampsia delivered a live male baby but shifted to NICU i/v/o antenatally diagnosed hypoplastic left heart syndrome. baby was discharged on 8th day of life. On 21st day of life baby presented with cold peripheries and gasping. Chest x-ray showed bilateral infilteration and baby succumbed due to severe pneumonia and cardiac arrest.

Keywords: congenital heart disease, hypoplastic left heart syndrome, cesarean section, pre-eclampsia, cardiologists

Introduction
Hypoplastic left heart syndrome [HLHS] is a congenital heart disease which has multiple etiological factors. This syndrome was first described by Lev in 1952 [3], later it is named by Noonan and Nadas in 1958 [4]. This syndrome represents in 2-9% of congenital heart disease [5)]. It is characterized by underdevelopment or absence of the left ventricle. Features of HLHS include varying degrees of hypoplasia of left ventricle, mitral valve and aortic valve atresia or stenosis, and hypoplasia of the ascending aorta. According to Barber G *et al*. coarctation of the aorta may be associated in 67% to 80% of cases [6]. It occurs predominantly in males [7]. It also has genetic predisposition. Most common etiology is idiopathic. This syndrome can be diagnosed by anomaly scan and confirmed by fetal echocardiography.

Case Report
A Case of primigravida with 35week period of gestation with left hypoplastic heart syndrome with single umbilical artery (detected at 24 weeks of gestation on ultrasonography-anomaly scan, cause for the anomaly is not known) was coming for regular antenatal checkups. Married life -6years, non-consanguineous marriage, no signicant family history is noted. antenatal counselling of hypoplastic left heart syndrome like chances of neurodevelopmental problems (30%), dysmorphic features (in 40% of cases), need for emergency surgery immediately after birth which is associated with improving survival rates (90-95%), higher rates of morbidity and mortality in untreated babies who often succumb during first or second week of life, has been explained to the parents and they wanted to continue the pregnancy. Patient came with c/o headache and vomiting since 1 day. On examination no pallor, no pedal edema, pulse rate-82/min, BP was 160/100 mmhg, per abdomen examination-Epigastric tenderness +, uterus 36weeks, irritable, cephalic presentation, fetal heart rate good. P/V cervical partially effaced, os 2cm dilated, bag of membranes forming, vertex -2 station, pelvis adequate. she was started with antihypertensives in view of impending eclampsia Emergency cesarean section done, delivered a live male baby cried immediately after birth (APGAR 1"-7/10, 5"-8/10) but shifted to NICU i/v/o antenatally diagnosed hypoplastic left heart syndrome. Mother condition was good and suture removal done on post op day 6 and was discharged on post op day 7. At NICU, baby was given prostaglandin infusion for 3 days. 2D ECHO showed hypoplastic left heart syndrome with aorta and mitral atresia, hypoplastic aorta and transverse arch, severe pulmonary hypertension. Parents are advised for immediate surgical intervention but post-surgical outcome was poor as this condition was associated with severe pulmonary hypertension. So, parents denied the procedure. The baby was given palliative treatment and discharged on 8th day of life in a stable condition.
Baby was brought to our hospital on 21st day of life with cyanosis, cold peripheries and gasping. Baby was taken to NICU intubated immediately. chest x-ray showed bilateral infilteration with right sided consolidation. Despite of all resuscitative efforts, baby succumbed. Cause of death cardiac arrest secondary to hypoplastic left heart syndrome with severe pulmonary hypertension with severe pneumonia with shock.

Discussion
Hypoplastic left heart syndrome is abnormal development of left sided cardiac structure, leading to obstruction to blood flow from

the left ventricular outflow tract. It includes underdevelopment of left ventricle, aorta and aortic arch and mitral atresia or stenosis.

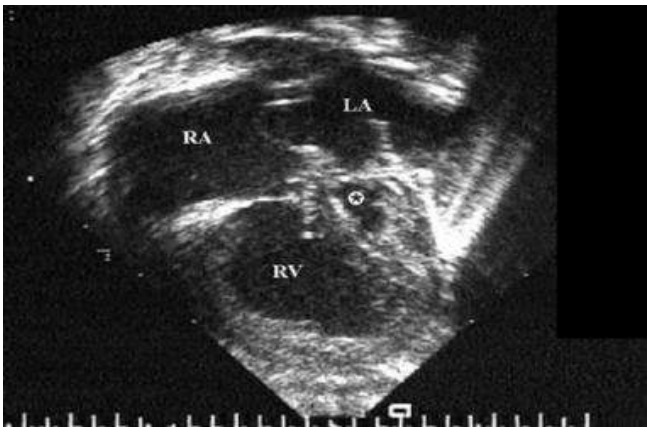

Fig 1

USG Showing Abnormalities in HLHS

This syndrome can be diagnosed by anomaly scan and confirmed by fetal echocardiography and furthur evaluation may include genetic testing. Prenatal diagnosis helps mainly in counselling the parents about the prognosis of the disease and plan for delivery in tertiary centre.

Pathophysiology

As soon as the baby is delivered, normal physiological changes occurs which leads to lethal hemodynamic disturbances in HLHS neonate [8]. The three major hemodynamic changes occur are gradual decrease in pulmonary vascular resistance, spontaneous constriction of ductus arteriosus and inadequacy of the interatrial connection [9]. Coarctation of the aorta may impede retrograde blood flow to a diminutive ascending aorta and thus decrease the adequacy of coronary blood flow [10, 11]. Because of all these changes systemic and coronary blood flow decreases which leads to tissue hypoxia, metabolic acidosis furthur which leads to vascular shock and death [9, 10].

Clinical Features

Most of the babies present with respiratory distress, tachypnea and mild cyanosis, shock and less commonly with severe cyanosis between 1-3 days of life [9]. Survival rate is only 4.5 days as it is associated with 95% mortality. Most infants die with in first two weeks of life [9, 11, 12]. Some infants will survive beyond 60 days without any surgical intervention but there will be a development of pulmonary hypertension [11, 13]. This condition may be isolated or associated with known genetic syndromes in 5-15% of infants, like turner syndrome, trisomy 13, 18, or 21, jocobsen syndrome (11q deletion), holt oram syndrome, Rubinstein-Taybi syndrome. Occasionally it is familial and inherited as an autosomal recessive trait.

Diagnosis

Antenatally detected by fetal 2D ECHO. Chest radiograph shows cardiomegaly with increased pulmonary vascularity. Initial ECG may be normal but later shows prominent p waves and right ventricular hypertrophy. The echocardiogram is diagnostic that demonstrates absence or hypoplasia of mitral valve and aortic root, a variaibly small left atrium and left ventricle and a large right atrium and right ventricle. Pulmonary vessels are assessed by color flow Doppler studies. Others are Doppler echo, angiography.

Management

Medical management is difficult if these babies survive. Some medical management has been studied using ACE inhibitors but no benefits are seen [14]. Ideal medical management for this condition remains inconclusive.

Surgical management includes traditional surgical staged palliation which consists of norwood procedure at birth, a stage II superior cavopulmonary connection, generally performed at 4-6 months of age, and a completion fontan at 18-48 months of age. Now recently hybrid procedure has been proposed instead of initial norwood procedure as hybrid is less invasive but physiology remains same in both norwood and hybrid procedure. perioperative survival rate for norwood operation is 47-85%. Recently, there has been renewed enthusiasm for right ventricle to pulmonary artery shunt (RVPAS) as a source of pulmonary blood flow for the norwood procedure. But disadvantage of this is need to perform a ventriculotomy, with the potential risk to ventricular function and arrhythmia generation. Predictions made that cardiac transplantation would be the final option. Babies who survive with stage 1 palliation may not be suitable for further procedures, for these infants cardiac transplantation may be considered but rejection is the major cause of death after transplantation [15].

Prognosis

Untreated babies most often succumb during first few months of life. Upto 30% of infants have major or minor central nervous system abnormality. Other dysmorphic features in upto 40% infants. Thus, careful preoperative evaluation (genetic, neurologic, ophthalmologic) should be performed. Whether the poor neurodevelopmental outcome is due to genetically associated central nervous system malformation, prenatal central nervous system injury, alterations of cerebral hemodynamics during bypass surgery, or poor post-operative perfusion is unknown. poor outcome is associated with prematurity, chromosome syndromes and poverty [16].

Conclusion

Once the diagnosis is confirmed, counselling is necessary. Options are optimal therapy i.e from no intervention to surgical thereapy. Palliative or comfort care is ideal option for many parents. With palliative therapy the long-term immunosuppression is possible. side effect leads to high operative mortality and also affects the quality of life. Cardiac transplantation has got high operative survival but associated with severe rejection.

For this type of cases, termination of pregnancy is advisable.

References

1. Morris CD, Samanek M, *et al*. Hypoplastic left heart syndrome: natural history in a geographically defined population**.** Pediatrics. 1990; 85(6):977-983.
2. Congenital heart disease among 815, 569 children born between 1980 and 1990 and their 15-year survival: A

prospective Bohemia survival study. Pediatr cardiol. 1999; 20(6):411-417.
3. Chameides L, Brownell LG, *et al*. The Regional Infant Cardiac Program. A six-year evaluation**.** Conn Med. 1975; 39(11):709-710.
4. Inheritance of hypoplastic left heart syndrome (HLHS): further observations**.** Clin Genet. 1976; 9(2):245-249.
5. Lev M. Pathologic anatomy and interrelationship of hypoplasia of the aortic tract complexes. Lab Invest. 1952; 1:61-70.
6. Noonan JA, Nadas AS. The hypoplastic left heart syndrome: an analysis of 101 cases. Pediatr Clin North Am. 1958; 5:1029-56.
7. Barber G. Hypoplastic left heart syndrome. In: Garson A, Bricker JT, Fisher DJ, Neish SR, eds. The Science and Practice of Pediatric Cardiology, 2nd edn. Volume II. Baltimore: Williams and Wilkins. Report of the New England Regional Infant Cardiac Program, Pediatrics. 1980; 65(Suppl):375-461.
8. Elzenga NJ, Jonas RA, *et al* Coarctation and related aortic arch anomalies in hypoplastic left heart syndrome. J Thorac Cardiovasc Surg. 1985; 8:379-89.
9. First-stage palliation of hypoplastic left heart syndrome. J Thorac Cardiovasc Surg. 1986; 92:6-13.
10. Perloff JK. The hypoplastic left heart. In: Perloff JK, ed. The Clinical Recognition of Congenital Heart Disease, 4th edn. Philadelphia: WB Saunders Co, 1994, 727-37.
11. Cohen DM, Allen HD. New developments in the treatment of hypoplastic left heart syndrome. Curr Opin Cardiol. 1997; 12:44-50.
12. Rosenthal A. Physiology, diagnosis and clinical profile of the hypoplastic left heart syndrome. Progress Ped Cardiol. 1996; 5:19-22.
13. 10.Charpie JR, Kulik TJ. Pre- and post-operative management of infants with hypoplastic left heart syndrome. Progress Ped Cardiol. 1996; 5:49-56.
14. Hoshino K, Ogawa K, Hishitani T, Kitawaza R, Uehara R. Hypoplastic left heart syndrome: Duration of survival without surgical intervention. Am Heart J. 1999; 137:535-42.
15. Jacobs ML, Blackstone EH, Bailey LL. Intermediate survival in neonates with aortic atresia: a multi-institutional study. J Thorac Cardiovasc Surg. 1998; 116:417-31.
16. Morris CD, Outcalt J, Menashe VD. Hypoplastic left heart syndrome: natural history in a geographically defined population. Pediatrics. 1990; 85:977-1000.
17. Hsu DT, Zak V, Mahony L, Sleeper LA, Altmann K,Ghanayem NS, Margossian R, Chung WK, *et al*; Pediatric Heart Network Investigators. Enalapril in infants with single ventricle: results of a multi-center randomized trial. Circulation. 2010; 122:333:340. doi:10.1161/CIRCULATIONAHA.109.927988.
18. Razzouk AJ, Chinnock RE, Gundry SR, Bailey LL. Cardiac transplantation for infants with hypoplastic left heart syndrome. Progress Ped Cardiol. 1996; 5:37-47.
19. Kliegman, ST Geme, Blum, Shah, Tasker, Wilson *et al* editors, Nelson textbook of pediatrics. 21st ed. Elsevier. Chapter; 458.10:9521-24.