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Asymmetrical Septal Hypertrophy: A Rare Case of Respiratory Distress in an Infant of a Diabetic Mother

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Abstract **Case Report**

Cardiomyopathy is a disease that affects the myocardium and causes mechanical or electric dysfunction. Hypertrophic cardiomyopathy comprises various cardiac components such as thickening of ventricular walls, hypertrophy of interventricular septum thereby resulting in subaortic stenosis, systolic and/or diastolic dysfunction. This cardiac complication is a known but rare entity in infants of diabetic mothers wherein the clinical presentation ranges from being asymptomatic to respiratory distress to congestive cardiac failure. Incidence of Congenital Heart Disease in infants of Diabetic Mothers is 5% and the percentage of symptomatic HOCM babies is 12%. Here, we report a term neonate, born to a $G_2P_2L_2$ mother with Gestational Diabetes Mellitus with poor glycemic control, presenting with respiratory distress manifesting as silent tachypnea at 24 hours of life, diagnosed as Asymmetrical Septal Hypertrophy on 2D-Echo, treated successfully with Propranolol and showed resolution of hypertrophy in follow-up echocardiography. In this condition, foetal hyperinsulinemia is responsible for an increase in synthesis and deposition of fat and glycogen in the myocardial cells which explains the cardiac hypertrophy. Due to increase in the thickness of the Interventricular Septum, there is Left Ventricular Hypertrophy and during systole, the systolic anterior motion of the anterior leaflet (SAM) of the aortic valve is elongated. Therefore, due to this obstruction of the orifice by the aortic valve, there is reversal of blood flow clinically manifesting as Silent Tachypnea. Symptomatic babies are treated with Oral Propranolol which acts by improving left ventricular outflow obstruction.

Keywords: Hypertrophic Obstructive Cardiomyopathy, Infant of Diabetic Mother, Septal Hypertrophy, Silent Tachypnea, Foetal Hyperinsulinemia.

INTRODUCTION

Cardiomyopathy is a disease that affects the myocardium and causes mechanical or electric dysfunction. Hypertrophic cardiomyopathy comprises various cardiac components such as thickening of ventricular walls, hypertrophy of interventricular septum thereby resulting in subaortic stenosis, systolic and/or diastolic dysfunction.

We report a neonate that was diagnosed as a case of asymmetrical septal hypertrophy born to a mother with Gestational Diabetes Mellitus (GDM) with poor glycemic control, presenting with respiratory distress, treated successfully with oral Propranolol, and showed resolution of hypertrophy in follow-up transthoracic 2-D echocardiography.

GDM is a frequent complication seen in pregnancy with incidence in the increasing trend recorded in developing countries. Diabetes in pregnancy results in an increased risk of foetal, neonatal, and long-term complications in the newborn (listed below).

Congenital – Neural tube defects, Intestinal anomalies, Cleft palate, Skeletal Malformations, Flexion Contractures.

Respiratory – Organogenesis, Transient Tachypnea of Newborn (TTNB), Respiratory Distress Syndrome (RDS), Pneumothorax.

Cardiac – Organogenesis, Hypertrophic Obstructive Cardiomyopathy (HOCM), Atrial Septal Defect (ASD), Ventricular Septal Defect (VSD), Transposition of Great Arteries (TGA), Patent Ductus Arteriosus (PDA).

Metabolic – Hypoglycemia, Hypocalcemia, Hypomagnesaemia

Others – Hyperbilirubinemia, Polycythemia, Macrosomia, Prematurity [1, 2]

Incidence of Congenital Heart Disease in normal newborns is 0.8% whereas in Infants of Diabetic Mothers (IDM), it is as high as 5%. The incidence of Asymptomatic HOCM is 36% while the percentage of symptomatic babies is 12% [3].

When routinely searched for with an echocardiographic scan HOCM is found in 30% of IDM, while symptomatic HOCM occurs in 12.1%[11] The left ventricular mass and contractility are increased and there is left ventricular outflow tract (LVOT) obstruction with apposition of the anterior leaflet of the mitral valve to the interventricular septum during systole. Cardiac output is significantly reduced, secondary to reduced stroke volume and is directly related to the degree of septal hypertrophy [12].

This asymmetric septal enlargement, with a disproportionately hypertrophic septum, is an anabolic result of foetal hyperinsulinemia triggered by maternal hyperglycemia during the third trimester. Cardiac septum hypertrophy correlates with maternal glycosylated haemoglobin levels and high levels of foetal insulin better than with macrosomia.

The severity of IDM cardiomyopathy can vary from an incidental finding on echocardiography (30% of cases) to an infant with severe symptoms of congestive heart failure (12% of cases) [13].

CASE REPORT

A term (39-weeks period of gestation) male baby, with birth weight of 3.97 kg was born to a 26-year-old $G_2P_2L_2$ mother via Lower segment Caesarean Section (LSCS) in view of Premature rupture of membranes (PROM). Baby had a successful transition from intrauterine to extrauterine life with APGAR scores of 8/9 at 1/5 minutes respectively. The mother was diagnosed with GDM in the 28th week, was on subcutaneous Insulin treatment but maintaining poor glycemic control. There was no other significant antenatal history with all routine scans revealing no abnormalities. Baby was monitored for sugars and respiratory distress and in the normalcy of both, was shifted to mother's side after 2 hours of observation in the Neonatal Intensive Care Unit (NICU).

Physical examination of the baby showed a macrosomic infant (weight >90% percentile for age), thick skin folds, broad shoulders, and a plethoric look. No other obvious malformations were noted.

At 24 hours of life, the baby was brought to the NICU with complaints of poor feeding, and an increased work of breathing. On examination, there was only tachypnea with respiratory rate of 82/min and no other significant findings. Provisionals of TTNB, Aspiration Pneumonia, Sepsis, Asymmetrical Septal Hypertrophy and other Congenital Heart Diseases were kept in mind and the baby started on oxygen support via nasal prongs at 1L/min thereafter maintaining saturations >93%.

Baby was investigated with a sepsis screen of Complete Blood Picture (CBP), C-reactive Protein (CRP) and Blood Culture/Sensitivity, Chest X-Ray (CXR) and Serum Calcium, which was in the favour of sepsis (CRP = 29.53, White Blood Cells= 25,000), Hypocalcemia (7.2mg%), CXR was done which showed cardiomegaly (Cardio-Thoracic ratio of 0.8) and plethoric lung fields. Baby was started on antibiotics for sepsis control, calcium supplementation initiated, and 2-Dimensional Transthoracic Cardiac Echocardiography scheduled to investigate the cause of cardiomegaly. The Echocardiography was suggestive of marked Inter Ventricular Septal Hypertrophy with significant reduction in volume of ventricular cavity i.e., features pointing towards Asymmetrical Septal Hypertrophy under HOCM.

Baby was started on Oral Propranolol at 2 mg/kg/day 12th hourly with regular monitoring of sugar, heart rate and blood pressure. Distress gradually subsided over the next 24-36 hours. Over the next 2 days the baby was weaned off Oxygen support, oral feeds initiated and monitored for 2 more days and discharged on oral Propranolol. Follow-up was scheduled at 2 weeks of age at the time of which tapering of dose was started and eventually stopped at 4 weeks of age.

The repeat transthoracic echocardiography done at 6 months of age showed resolution of hypertrophy.

DISCUSSION

The diagnosis of sepsis would explain the distress but not the cardiomegaly. There were no associated signs of rales, cyanosis, and hepatomegaly to give congenital heart disease (CHD) as the diagnosis leaving the only significant symptom the baby presented with, the silent tachypnea, wherein the level of tachypnea does not match the severity of the retractions which in this baby's case, was completely absent.

Gestational Diabetes Mellitus is defined as carbohydrate intolerance of variable severity first diagnosed in pregnancy. Complications of late onset GDM include Somatohypertrophy, Metabolic, Hyperbilirubinemia, Polycythemia, Macrosomia and Birth Trauma [1].

The mechanism of Foetal Hypoglycemia is explained by the Pederson Maternal Hyperglycemia-Foetal Hyperinsulinemia Hypothesis which states: Maternal Hyperglycemia leads to Foetal Hyperglycemia which in turn causes Foetal Hyperinsulinemia resulting in Foetal Hypoglycemia. The Foetal Hyperinsulinemia is responsible for an increase in synthesis and deposition of fat and glycogen in the myocardial cells which explains the cardiac hypertrophy [4, 5].

Due to increase in the thickness of the Interventricular Septum, (>1.3 times the normal thickness is significant) there is Left Ventricular Hypertrophy and during systole, the systolic anterior motion of the anterior leaflet (SAM) of the Aortic Valve is elongated. Therefore, due to this obstruction of the orifice by the Aortic Valve, there is reversal of blood flow clinically manifesting as silent tachypnea [6, 7].

Inotropic Agents like Dopamine and Dobutamine are contraindicated as they end up reducing the ventricular size and thereby contribute to obstruction of cardiac outflow. Beta Blockers cause reduction in maximal contraction velocity leading to change in Systolic Anterior Motion and thereby improving left ventricular outflow obstruction [8].

Studies similar to ours: Codazzi, A.C., Ippolito, R., Novara, C. *et al.* [9] observed similar results with use of Propranolol in two babies born to diabetic mothers and so did the study of Sharma D, Pandita A, Shastri S, Sharma P. who also documented resolution of septal hypertrophy by 6 months with use of Propranolol [10].

CONCLUSION

Uncontrolled Diabetes Mellitus in mothers has proven to have detrimental side effects for the newborn making them a high-risk infant. Smooth, uneventful transition to extrauterine life, close monitoring in the immediate postnatal period, early diagnosis of underlying abnormality and periodic screening and follow-ups for associated congenital anomalies ensure that these babies are well taken care of. Beta Blockers must be started for symptomatic babies like this one but strictly monitored for side effects like bradycardia and hypoglycemia.

Septal Hypertrophy is one of the rarest manifestations of HOCM with Incidence of Congenital Heart Disease in normal newborns at 0.8% whereas in Infants of Diabetic Mothers, it is as high as 5%.

Marked Inter Ventricular Septal Hypertrophy with significant reduction in volume of ventricular cavity. Baby was started on Oral Propranolol at 2 mg/kg/day 12th hourly with regular monitoring of sugar, heart rate and blood pressure. Distress gradually subsided over the next 24-36 hours.

Follow-up was scheduled at 2 weeks of age at the time of which tapering of dose was started and eventually stopped at 4 weeks of age. The repeat Echo done at 6 months of age showed resolution of hypertrophy.

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