Persistent Pulmonary Hypertension of the Newborn in donor twin: Another complication of fetal anemia in twin to twin transfusion syndrome?

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Citation

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Abstract

Twin to twin transfusion syndrome (TTTS) is a major complication of monochorionic multiple gestation associated with significant perinatal morbidity and high mortality rates. Here we report a case of such morbidity, persistent pulmonary hypertension of the newborn (PPHN) in a near term donor twin probably secondary to significant anemia, needing rescue therapy with high frequency oscillatory ventilation (HFOV) and inhaled Nitric oxide (INO). As there has been no documented case of PPHN in the donor twin of
newborns with TTTS and given the risk of TTTS and prevalence of preterm delivery, fetal and neonatal anemia presenting with PPHN should be considered in the immediate neonatal period, our case demonstrates that, with early and proper identification and aggressive management, successful outcome is possible in this group of neonates.

INTRODUCTION

Twin twin transfusion syndrome (TTTS) is a major complication of monochorionic multiple gestations associated with significant perinatal morbidity and high mortality rates. The incidence of TTTS has been reported as approximately 5.5 to 17.5% of all monochorionic pregnancies([1]). Although more than a century has passed since it was first described by Schatz et al in 1882 ([2]), the pathogenesis remains uncertain. Substantial progress has been made to understand the alternate possible pathophysiological mechanisms apart from the traditional transfusion theory. These theories backed by substantial basic research, range from loss of proteins from donor into recipient causing changes in osmotic pressure with fluid transfer over the placenta ([3]) increased atriopeptin and decreased antidiuretic hormone in the recipient ([4],[5]), to growth stimuli from the donor promoting growth of the recipient twin ([6]). Whatever the etiology is, the presence of functional placental vascular anastomoses is an essential condition for the development of TTTS ([7]). Here we report a case of persistent pulmonary hypertension of the newborn (PPHN) in a near term donor twin probably secondary to significant anemia, needing rescue therapy with high frequency oscillatory ventilation (HFOV) and inhaled Nitric oxide (INO).

CASE REPORT

The patient is a 1935gm female infant, second of the monochorionic diamniotic twins born by cesarean section at 35+2 weeks to a 28year old primiparous mother. The maternal history and routine pregnancy screening laboratory results were reported to be within normal limits. Her regular medications included prenatal vitamins, folic acid and calcium. Pregnancy was closely monitored from the first trimester and progressed without complications until she presented in preterm labor at 32+5 weeks. Ultrasound revealed monochorionic diamniotic twins, both in vertex position with a 14% discordance between their estimated fetal weights (there was no growth discordance on the ultrasound at 28 weeks), no other findings of TTTS were documented. She was treated with tocolytics; initially magnesium sulfate, subsequently terbutaline and nifedipine until 35weeks when tocolysis was discontinued. She received a course of antenatal corticosteroids, was treated with Azithromycin and then Amoxicillin- clavulanic acid for sinusitis. There were no signs of chorioamnionitis and gram stain of the amniotic fluid was negative. She remained hospitalized undergoing biweekly Non stress tests (NST's). At 35+2 weeks she again experienced preterm labor with

non reassuring fetal heart tracing and poor biophysical profile of 4 out of 10 of this twin (minus 2 for NST, minus 2 for breathing and minus 2 for fluid pocket) prompting delivery by emergency cesarean section. The baby, on delivery was pale and floppy with a heart rate under 100beats/min. She displayed minimal respiratory effort and was given positive pressure ventilation via Neopuff for one minute with improvement, switched to CPAP and transferred to the NICU. Apgar scores were 3 and 8 at 1 and 5 minutes respectively, her birth weight was 1935gms (20% discordance with twin A weighing 2440gms), venous cord pH was 7.3 with a base deficit of 3.7, arterial cord pH was 7.17 with a base deficit of 7.9 and hemoglobin (Hb) was 5.1g/dl (twin A's Hb was 23.4gm/dl i.e. >5gm/dl). Initial chest radiographs showed features consistent with mild respiratory distress syndrome, retained lung fluid with a small right sided pleural effusion.

Her immediate postnatal course was complicated by multiple desaturations and hemodynamic failure necessitating intermittent positive pressure mechanical ventilation, surfactant replacement, a total of 50cc/kg of leukocyte reduced packed red blood cell transfusion and triple inotropic support of Dopamine , Dobutamine and Epinephrine. Despite above measures and support oxygenation only marginally improved and ventilation remained poor, at 16hrs of life she was placed on high frequency oscillatory ventilation and Nitric oxide soon afterwards for worsening oxygenation and oxygenation indices (OI) secondary to PPHN. Echocardiogram done within 24 hours of life showed equal left and right ventricular pressures with tricuspid regurgitation and bidirectional flow across the patent ductus arteriosus consistent with PPHN. Oxygenation and ventilation improved and she was weaned off nitric oxide on day 5 and inotropes on day 7. She was further weaned to room air by day 11, on full enteral feeds by day 15 and discharged home after three weeks of hospitalization.

Placental pathology showed monochorionic placenta with disrupted and indeterminate membranes. The maternal surface had a line demarcating the markedly congested half of twin A from the markedly pale half of twin B.

DISCUSSION

Although substantial progress has been made in the early diagnosis and subsequent aggressive antenatal management with serial amniocentesis and laser ablation of chorionic vessels joining the two circulations, TTTS continues to be associated with significant perinatal morbidity and mortality. Perinatal morbidity is generally related to complications of prematurity and specifically to the donor and the recipient circulatory status. In our case classic antenatal features of chronic TTTS were absent, but an antenatal weight discordance of 14%, oligohydramnios at 35weeks associated with anemia and hemodynamic compromise in this twin at birth suggests an acute on chronic form of TTTS or an acute form of TTTS complicating intrauterine growth restriction of this twin. Blickstein et al ([8]) reported an acute form of TTTS which can occur during labor or during delivery of the first twin. Physical findings of circulatory insufficiency such as tachycardia, pallor, poor peripheral circulation and hypotension as a result of hypovolemia in the donor twin, requiring immediate volume expansion with packed red blood cells has been reported in the literature([9]). The recipient twin may be at risk of developing polycythemia-hyperviscosity syndrome which can be managed by partial exchange transfusion and correction of electrolyte abnormalities. Finally TTTS can predispose both donor and recipient to the development of nonimmune hydrops fetalis which may require intensive cardiorespiratory support and monitoring([10]).The pathophysiology of fetal anemia leading to PPHN in immediate newborn period is uncertain. PPHN, as a consequence of marked anemia secondary to fetomaternal hemorrhage has been reported([11],[12],[13]).

The mechanisms involved in pulmonary endothelial adaptation to sustained hypoxia includes a reduction in nitric oxide production, possibly through reduced activity of nitric oxide synthase which is mainly responsible for initial transition from fetal circulation in the newborn ([14]). Additionally, Anderson et al ([15]) reported that hypovolemic, anemic hypoxia is less well tolerated than hypoxic hypoxia and cardiac output and hemoglobin are the essential elements in determining systemic oxygen transport to facilitate normal transition in the newborn period. Although no other documented case of PPHN in the donor twin of newborns with TTTS has been reported, our case demonstrates that the risk is real, and that, more importantly, the response to conventional treatment with immediate volume expansion, inotropic support and adequate oxygenation with available ventilation is effective. Given the risk of TTTS and prevalence of preterm delivery, fetal and neonatal anemia presenting with PPHN should be considered in the immediate neonatal period. Our case demonstrates that, with early and proper identification and aggressive management, successful outcome is possible in this group of

neonates.

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