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Immunologic and Nonimmunologic Hydrops Fetalis

(Report of Two Surviving Cases).

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Introduction

An association between Rhesus immunization and hydrops fetalis is well established.[1,2] However, there are many other conditions that can result in the clinical condition of hydrops fetalis, for example, alpha-thalassemia,[3] multiple congenital abnormalities,[4] placental transfusion syndrome in twins,[5] pregnancy complicated with toxemia,[6] hamartoma of lung,[7] hemangioendothelioma,[8] neuroblastoma,[9] hydrops fetalis due to Coxsackie virus B3 pancarditis,[10] congenital syphilis,[11] congenital hepatitis, congenital nephrosis, renal-vein thrombosis, chorionic-vein thrombosis, umbilical-vein thrombosis, and finally no apparent cause (idiopathic).[12,13] These are so called nonimmunologic hydrops fetalis. Pathogenesis of nonimmunologic hydrops fetalis is still poorly understood.[14,15]

The purpose of this paper is to report two cases of surviving hydrops fetalis. One of these cases was diagnosed as hydrops fetalis due to Rh isoimmunization and the other as being due to placental transfusion syndrome in twins. The rarity of survival of these cases prompted us to record our experience with these patients.

Case I. The mother was a 33 year old, gravidal, para O, who had an uneventful preganancy until the 29 th week of gestation. During an examination because of right upper quadrant pain, it was noted that there was no fetal heart beat in one of the twins. Due to this diagnosis of intrauterine death, a Caeseran section was performed at the 30 th week of gestation

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at another hospital. Twin A was a slightly macerated, deep purple colored female infant, and Twin B was a hydropic female with an apgar score of three. Her condition improved after five minutes of resuscitation. A postmortem examination of Twin A revealed a macerated stillborn plethoric infant with congestion in all the organs.

Twin B was transferred to the Hacettepe Premature Unit. Upon admission she was very pale, mimicking hydrops fetalis. Her birth weight was 1, 890 gm. The direct Coombs test was negative, her blood group was 0 Rh positive, the reticulocyte count was 11.2 per cent and she had many normoblasts in her peripheral blood smear; Hb was low (11.16 gm/100 ml). There were rales at the base of both lungs due to congestive heart failure. Since Twin B had anemia and Twin A had a plethoric appearance, the former was immediately diagnosed as having placental transfusion syndrome.

The anemia was treated by packed cell transfusion which the infant tolerated and responded to well. Within 24 hours the rales in both lung fields disappeared and child passed a large amount of urine. After this initial therapy she was put on elemental iron. She made good progress and was discharged at the age of 47 days in excellent condition with a Hb of 12.5 gm/100 and a 1.8 per cent reticulocyte count.

Case II. This was a male infant born to a gravida 2, para 1 mother after 39 weeks of gestation. The first child is a 6 year old boy in good health. There was no history of jaundice in the neonatal period.

Because the mother's blood group was AB Rh negative, a determination of anti Rh antibody titration was made at 38 weeks of gestation and it was found to be $\frac{1}{32}$ Saline and $\frac{1}{32}$ Bovine, positive. An amniosynthesis was made and spectro-photometric examination of the fluid revealed a large deviation from normal in optical density at 450 mp, indicating that it was in the area of the 3rd Liley zone. Because of this finding, three attempts were made to induce labor artificially, but all failed. The baby was delivered by Cesarean section.

The baby at birth weighed 3100 gm and looked extremely pale. There was generalized oedema. His abdomen was distended and examina tion showed that there were free ascites. He had rales in both lung fields. The liver was 5 cm below the right costal margin. The spleen was not palpable.

Laboratory tests: Cord blood examination revealed that Hb was 6.12 gm/100. The blood group of the baby was B Rh positive and Direct Coombs test was positive. Cord bilirubin level was 7.5 mg/100 ml.

Because of the baby's severe anemia, tachycardia and tachpnea, cyanosis, enlarged liver and generalized oedema, it was felt that the baby had heart failure. For this reason the baby was digitalized with cedilanid and also 0.1 ml of diuretic was given. A parasynthesis was made and a 150 cc fluid was removed from his abdomen immediately. As soon as the baby's condition improved, an exchange transfusion was performed with B Rh negative blood, with no complications.

The baby's condition improved. When he was two days old a parasynthesis was repeated and another 60 cc's of fluid was removed. Following this he had a total of six exchange transfusions, given according to his bilirubin levels.

On the 30 th hospital day, his Hb dropped to 5 mg/100 ml. Because of the tendency to have heart failure due to late anemia he was given a blood transfusion, with packed cells, and following this, his Hb rose to 8.3 gm, and the reticulocyte count was 1.8 %. The patient was discharged in good health.

Discussion

Treatment of the hydropic infant is rather difficult, since underlying causes of hydrops fetalis are varied. Sometimes prognosis is hopeless as in hydropic infants associated with multiple congenital abnormalities[4] and alpha-Thalassemi.[3]

In most cases, but not all, anemia is the most responsible trigger mechanism for the development of heart failure, and the clinical appearance of hydrops fetalis. If there is only anemia and heart failure present, prompt management of these babies is usually enough to save their lives as in our two cases. Literature has so far reported few surviving hydropic infants.[16, 17]

Peritoneal dialysis and positive pressure ventilation,[18] has been suggested as treatment for hydropic infant cases. O'Neill et al[19] suggested that extended exchange transfusion be used for these severely affected babies. Their aim of treatment at the early stage for hydropic infants is to bring the patient out of heart failure as quickly as possible by removal and replacement of blood by slow drip which can then be continued until the baby is completely out of heart failure. After correction of anemia and heart failure a full exchange transfusion can be done according to bilirubin levels of blood if necessary. Since the etiology of hydrops fetalis of one of our cases was anemia due to placental transfusion syndrome, we felt that several small packed cell transfusions were more preferable, as suggested by O'Neill et al.[19]

Our experience with these two cases of hydrops fetalis indicated that infants who are born with hydrops fetalis are worth trying to save, in spite of the high mortality rate associated with these cases.

Summary

Two surviving cases of hydrops fetalis are presented; the etiology treatment of these cases are discussed.

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