

LONG TERM OUTCOME OF INTRAUTERINE TRANSFUSIONS

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SUMMARY

In this article, we report a long term follow up of Turkey's first four cases of erythroblastosis fetalis who had been successfully treated with intrauterine transfusions between 1967-1969.

Key words : Erythroblastosis fetalis, Intrauterine transfusion.

INTRODUCTION

Although the wide clinical use of Anti D Immunoglobulin (Ig) since 1968 has significantly lowered the incidence of Rh-isoimmunization, especially in developing countries, cases sensitized with Rh-Antigen (Ag) continue to create many problems for obstetricians and neonatologists (1,2).

Intrauterine blood transfusion, which can be carried out in advanced perinatology centers since 1963, is an exciting approach in the treatment of erythroblastosis fetalis (3).

The purpose of this article is to present the 25 years follow up of 4 cases which underwent successful intrauterine intraperitoneal transfusion in 1967 at the Hacettepe University Children's Hospital.

CASE REPORTS

CASE 1

Baby D was born to a 27 year old Gravida 5 Para 2 B Rh (-) mother. Father's blood group was B Rh (+). Obstetric history revealed that the mother previously had a hydropic baby and a child with bilirubin encephalopathy.

The amniotic fluid readings of the mother were in the Liley zone 3 at 33 weeks of gestation, when an intrauterine intraperitoneal transfusion was

performed. A baby girl with a birth weight of 2670 gm was delivered with induction due to disturbances in fetal well-being monitoring at 34 and a half weeks. Her Apgar scores were 8 and 9 at 1 and 5 minutes respectively.

Physical examination revealed hepatosplenomegaly, abdominal distention and edema on the lower extremities. Baby's blood type was B Rh (+) and the direct Coombs test was positive. The cord blood hemoglobin (Hb) and bilirubin levels were 10.45 gr/dl and 3.3 mg/dl. Two exchange transfusions were performed on the 1st and 2nd postnatal days. Anemia was noted at 1 month of age. The patient was followed up to 25 years. All physical, neurodevelopmental and psychomotor examinations were found normal except bilateral sensorineural hearing loss detected on the audiologic examination at the age of two. She is now the top manager of a firm.

CASE 2

Baby B was born to a 29 year old Gravida 5 Para 2 A Rh (-) mother. Father's blood group was O Rh (+). Mother's prior pregnancy history revealed that her last three babies had died before birth because of severe hydrops fetalis. In her present pregnancy, amniotic fluid optic density readings were 0.161 at 31 week and 0.201 at 33 week corresponding to Liley zone 2 and 3 respectively. Two successful intrauterine intraperitoneal transfusions were performed at 31 and 33 weeks. A female infant was delivered at the 35 th gestational week with a birth weight of 2770 gm, and a 1 minute Apgar score of 9.

Physical examination revealed no pathological findings other than hepatosplenomegaly. Cord blood Hb and bilirubin levels were 16.3 gm/dl and 2.2 mg/dl respectively. The baby's blood type was A Rh (+) and direct Coombs test was (+). She had an uncomplicated clinical course and no exchange transfusion was needed.

She was found to have a severe late anemia associated with urinary tract infection at 2 months of age. She was followed up to 25 years and remained in good health physically and mentally. Neurologic development was also normal. She later graduated from the University of Istanbul, Faculty of Economics.

CASE 3

Baby F was born to a 38 year old Gravida 4 Para 2 B Rh (-) mother. Father's blood type was A Rh (+). The mother's last pregnancy had resulted in a postnatal death due to erythroblastosis fetalis. Amniotic fluid readings of the mother were found to be in the Liley zone 3 at 31 and a half gestational week. An intrauterine intraperitoneal transfusion was performed on the 32nd week. A female infant was delivered on the 34th gestational week with a birth weight of 1600 gr and a 1 minute Apgar score of 3, necessitating resuscitation. At birth the baby was in congestive heart failure and looked severely affected. Cord blood Hb was 7.30 gr/dl and bilirubin 10 mg/dl. Her blood group was B Rh (+) and Coomb's test was positive. An exchange transfusion was given immediately after birth. Eight more exchange transfusions were required within 4 days. The baby was discharged at 58 days of age and followed up to 24 years. No physical or neurodevelopmental abnormalities were reported in her routine physical examinations. She graduated from the University of Ankara, Faculty of Political Sciences.

CASE 4

Baby N was born to a 25 year old A Rh (-) mother. Father's blood group was O Rh (+). The mother's second pregnancy had ended with an abortion. The third pregnancy resulted in an uncomplicated, term vaginal delivery of a baby with erythroblastosis fetalis. At this present pregnancy, amniotic fluid readings were in the Liley zone 3 at 28 week of gestation after which 3 intrauterine intraperitoneal exchange transfusions were performed at 28,30 and 32 weeks respectively.

A male infant was delivered weighing 2380 gm at 34 weeks of gestation with a one minute Apgar score of 4. Physical examination disclosed generalized edema and hepatosplenomegaly. Cord blood Hb was 7.85 mg/dl and bilirubin was 9.5 mg/dl.

The baby underwent two exchange transfusions. He also was followed until 23 years of age and was found to be completely healthy.

Liver function tests and Hepatitis B surface antigen detection test were performed in all four cases and reported to be normal.

DISCUSSION

The discovery and subsequent expanded use of anti-D human Ig has drastically reduced the incidence of

Rh isoimmunization from a previous high of one in 120 to 150 pregnancies (4) Unfortunately, it has not eliminated this condition entirely and women who are already sensitized will continue to become pregnant especially in underdeveloped countries (5). Reasons for the appearance of new cases of Rh isoimmunization include failure to administer RhoGAM to all, failure to give adequate quantities of RhoGAM following massive fetomaternal hemorrhages, sensitization prior to delivery in the second and third trimesters and sensitization to irregular RBC antigens (2,5).

First in 1956, Bevis pointed out the relationship between the quantity of bilirubin pigment in amniotic fluid and the severity of fetal erythroblastosis (6). The interpretation of data from amniocentesis was subsequently refined and this became an important prognostic guide. Later on in 1963, Liley successfully transfused a severely sensitized erythroblastotic infant in utero using the intraperitoneal method and in doing so, created a new vision in obstetric care (3,7).

Following his reported success, criteria for the selection of candidates, formulas to calculate the optimal amount of blood to infuse and schedules for the timing of transfusions and deliveries were devised and modified by a number of investigators (8).

However, the outcome of this procedure was found doubtful in hydropic fetuses since they seem to have an inadequate absorption of erythrocytes from the peritoneal cavity. More recently it has been reported that direct intravascular transfusion in utero also can improve the outcome in hydropic fetuses (9,10). This intervention was first performed by fetoscopic approach. Nowadays it is carried out under ultrasonic guidance and is superior to intraperitoneal intrauterine transfusion in many aspects (2,11,12). Today, it is well known that intrauterine transfusion is essential in the management of the severely affected erythroblastotic fetus. Experience with this method has indicated that when used appropriately by well-trained physicians at referral centers, it can offer the potential for survival and a normal life to an infant who would otherwise certainly die. Turner and colleagues reported in 1975 that about 50 % of 44 surviving infants were found to be normal when indices of physical health, intelligence and social maturity were measured (13). Later on, Hardyment analyzed the results of a series of 15 published studies between 1969 and 1977. This includes the data from the study by Turner et al. A total of 450 infants were studied at intervals varying from 1 month to 10 years in this literature review. 78.2 % of the infants were normal, whereas 16.7 % had minor and 5 % had major CNS signs (14). It should be noted that some of the infants in these studies had sustained severe hypoxic insults during their intrapartum or neonatal periods (14).

In this article, we report the long term follow-up of Turkey's first four cases of erythroblastosis fetalis,

who had been successfully treated with intrauterine transfusions and graduated from the university.

Our cases although few in number, showed consistent results with the literature and only one out of 4 nonhydropic babies revealed sensorineural hearing defect.

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