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Hydrops Foetalis

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Five cases of hydrops foetalis are reported. In four infants the condition was due to isoimmunization, in one infant in all probability to hydramnios of the mother. Of the five patients, two survived. Aetiology and treatment of hydrops foetalis are discussed.

Congenital hydrops foetalis is the most severe manifestation of neonatal haemolytic disease. In the great majority of cases it is induced by Rh isoimmunization. With the increasing application of anti-D IgG prophylaxis a decrease in the occurrence of neonatal haemolytic disease and hydrops may be expected. In addition, after successful intrauterine transfusions 42% of the hydropic newborns were delivered alive [6] and in such a condition that some of them could be saved by intensive obstetric and paediatric treatment. This is true for even the most severely damaged category, that of foetal and placental hydrops, which includes 8 to 10% of the infants of sensitized mothers.

Aetiology

Neonatal hydrops may be induced besides isoimmunization by a number of other factors. These are,
maternal diabetes [14];
severe maternal infections [2];
ABO incompatibility [10];
infrequent incompatibilities [10];
congenital nephrotic syndrome [14];
chronic foeto-maternal transfusion [5, 18];
hydramnios of mother [4];
congenital toxoplasmosis [17];
twin transfusion syndrome [14];
cytomegalovirus disease [5];
congenital syphilis [1];
alpha-thalassaemia [14];
glucose-6-phosphate dehydrogenase deficiency [15];
haemangioendothelioma [16];
pulmonary lymphangiectasis and congenital anomalies of the lungs [5];
thrombosis of umbilical vein [5];
foetal neuroblastoma [5];
achondroplasia [11];
Down's syndrome [5].

Pathomechanism

The pathomechanism of hydrops induced by isoimmunization is shown schematically in Fig. 1.

The process starts with anaemia, hypoproteinaemia and oedema

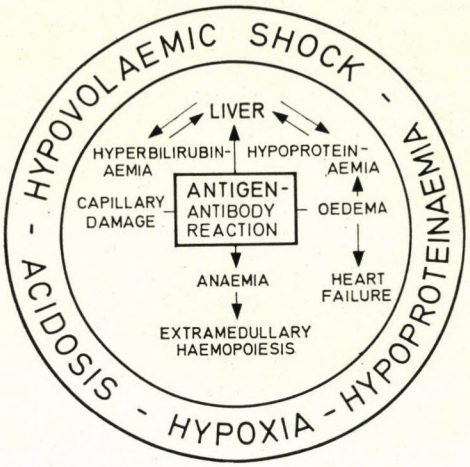

FIG. 1. Mechanism of Hydrops Foetalis

affects an increasing number of organ systems; finally, it takes a very rapid course. The lethal outcome can be only prevented by interrupting the vicious circle by a series of rapid interventions.

For long, hydrops foetalis was practically a postmortem diagnosis, there having been very few survivors. Therefore, the diagnostic criteria are mostly lacking in the literature. The leading symptom is oedema, which is more pronounced than that common in underweight newborns or in the infants of diabetic mothers. The presence of ascites is also characteristic. Accounting to the severity of the process, some additional symptoms are seen such as anaemia, excessive hypoproteinaemia, and a shock-like state. Death frequently occurs before the grave jaundice could have developed. Of these symptoms, hydrops and anaemia arise during intrauterine life.

REPORT OF CASES

In the period 1969 to 1972 we have observed five cases of hydrops foetalis among about 12,000 infants delivered in our hospital and those admitted for exchange transfusions (Table I).

Case No. 1. M. Gy., born from the third pregnancy of the 30-year-old mother. Her first pregnancy had been interrupted due to toxaemia. The second had resulted in a pre-term stillborn infant weighing 1500 g. During this third pregnancy she had had frequent vomitings. During the entire course of pregnancy the Galli-Mainini reaction was constantly negative. At the end of pregnancy a hydramnios had been observed. Delivery occurred in the 39th week of gestation and was normal. The blood group of both mother and infant was equally B Rh positive. Birth weight of the child was 2750 g, that of the placenta, 850

g; its diameter was 30 by 23 cm and its thickness, 3 cm. It was oedematous, meat-like, with extensive white necrotic areas (Fig. 2).

The newborn presenting with considerable oedema obtained a 1 minute Apgar score of 8. When he was transferred to us, at 10 min. the score was 2, as he failed to breathe and was cyanosed and atonic. All over the body marked oedema and some petechiae could be observed. The head was so oedematous that the eyes were invisible. The skin was pale without jaundice. The liver reached 4 cm below the costal margin, the spleen was just palpable. After cleaning the airways and resuscitation, spontaneous respiration started. On cutting the umbilical stump, blood escaped jetwise from the umbilical vein. Then 0 Rh negative blood, treated by soluble group substance (Witebsky) was administered in 20 ml portions; simultaneously, 10 ml doses of blood were removed. After the removal of 60 ml blood, respiration became periodical, heart sounds were almost inaudible, repeated attempts at resuscitation failed. Death occurred at three hours of age.

The following laboratory tests could be performed: Hb, 5.8 g per 100 ml; serum bilirubin, 0.31 mg per 100 ml; SGOT, 150 U; SGPT, 4 U; serum olechsterol, 147 mg per 100 ml; serum sodium, 138 mEq/1; serum chloride, 104 mEq/L; total serum protein: 3.4 g per 100 ml, with albumin 57%, $alpha_1$ globulin 7%, gamma globulin 5%. Wassermann test, negative. Immunoelectrophoresis revealed two prealbumins, with a considerably increased haptoglobin. Both IgA and IgM were markedly decreased. There was a pronounced increase of alpha- and beta lipoproteins. No incompatibility whatever could be detected even to infrequent sub-groups. Performance of further examinations was not permitted by the mother.

Case 3. L. L. The patient was born from the third pregnancy of the 28 years-old mother who has one living, healthy child and had one artificial abortion. Her blood group was AB Rh negative, that of the infant, B Rh positive. Indirect Coombs test was positive at a dilution of 1 to 32.

In the 36th week of pregnancy amniocentesis was performed. Spectrophotometry of the amniotic fluid indicated foetal damage, corresponding to the II/c region of Liley's scheme (borderline case between severe and moderately severe). Immediate Caesarian section was performed.

Both the foetus and the placenta were hydropic. The newborn was delivered in asphyxia. He was immediately intubated, the meconium containing fluid was removed by aspiration. After 10 minutes resuscitation, spontaneous respiration started. Then the infant was referred to us; during transport in the ambulance car oxygen was continuously administered. At admission, the baby's weight was 2000 g. The skin was greyish, anaemic and moderately jaundiced. Oedema was most pronounced on the head and face. Muscle tonicity was reduced. The heart rate was 120, the heart sounds were clear

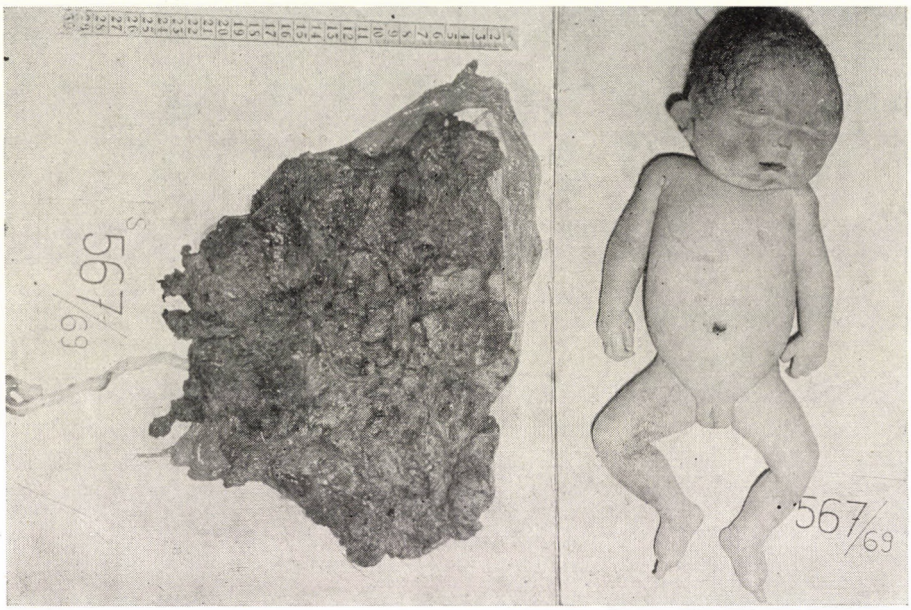

FIG. 2. Hydropic Newborn and Placenta

and rhythmical. Respiration rate was 38 per min, over the lungs rales were heard bilaterally. The abdomen was ballooning, ascites could be detected by percussion. The liver was massive, reaching 3 cm below the costal margin, the spleen was somewhat enlarged. Grasp reflexes could be elicited, the tendon reflexes were delayed.

At admission the serum bilirubin value was 8.9 mg per 100 ml, partly delayed direct, and partly indirect. The newborn was placed into the incubator, on an apnoe alarm device, and 40 vol. per cent oxygen was administered. A catheter was inserted into the umbilical vein; portal venous pressure was 9 to 10 cm H_2O. In two portions, altogether 70 ml red cell suspension treated with 0 Rh negative group substance was administered for the correction of anaemia. Simultaneously, prednisolone and cefaloridine treatment was started. At two hours, the haematocrit value was 30%, the serum bilirubin 13.0 mg per 100 ml. Abdominal puncture yielded 50 ml of yellowish ascitic fluid with a bilirubin content of 6.6 mg per 100 ml. At the age of 6 hours was the first exchange transfusion given, with 0 Rh negative group substance-treated blood under constant registration of heart function. The initial serum bilirubin value was 16.3 mg and, at the end of the transfusion, 7.9 mg per 100 ml. At the beginning, tachycardia, then later bradycardia was observed, with hypoxic signs on the ECG. The intervention was well tolerated, and soon diuresis started. At 19 hours the serum bilirubin was 12 mg per 100 ml, and Hb 13.7 g per

100 ml, the acid-base conditions were normal. The results of further laboratory tests are shown in Fig. 3.

After a repeated abdominal puncture which yielded 40 ml fluid, at the age of 27 hours a second exchange transfusion was given, as the serum bilirubin value had risen to 18.0 mg per 100 ml. By the end of the transfusion this decreased to 7.7 mg per 100 ml. Then it rose again and at the age of 72 hours it was 21.6 mg per 100 ml; therefore a third blood exchange was performed. At its end the serum bilirubin value was 8.2 mg per 100 ml. The considerable rebound observed during the first two exchange transfusions failed to appear after the third blood exchange.

On the third day petechiae appeared all over the body, the platelet count was 20,000 per μl. Simultaneously, the anaemia became marked, and had to be corrected by transfusions of fresh blood. Liver and spleen enlargement receded slowly and so did the serum bilirubin value with a gradual elevation of the direct fraction. With the increase of diuresis there was a visible decrease of the oedema; at 1 week, the infant weighed 1700 g. The SGOT value which ini-

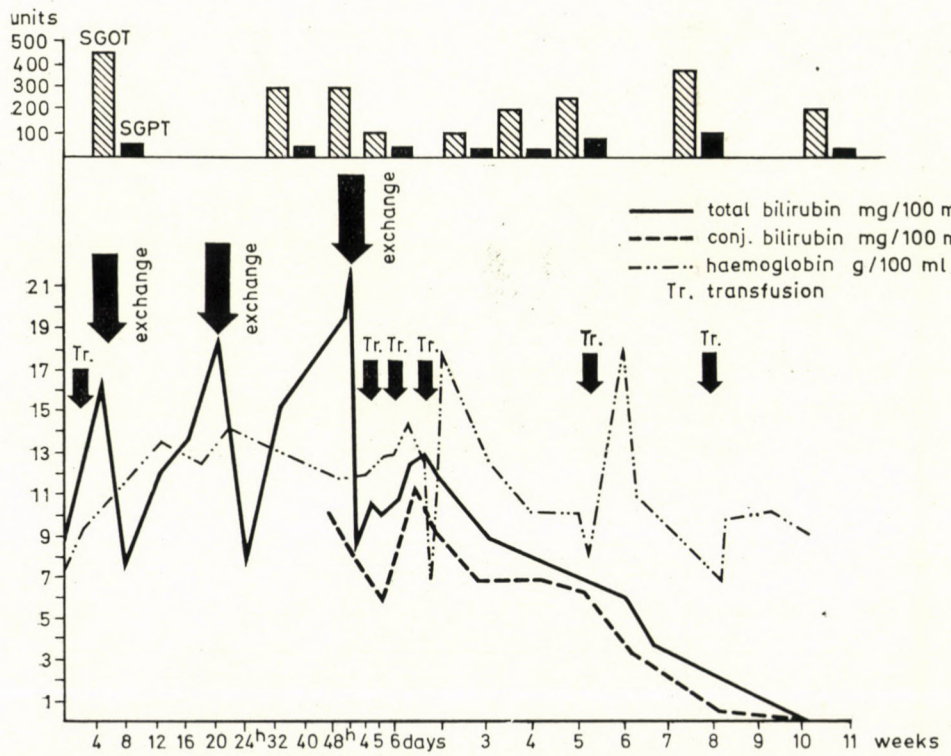

Fig. 3. Case No. 3.

tially was above 400 U, showed a transitory decrease, then a new increase to 150 U in the 7th week. From the 4th week on, the infant gained weight at a uniform rate, its condition was steadily improving, no neurologic involvement was apparent, only some liver and spleen enlargement persisted. He was discharged at 6 weeks with a weight of 3500 g.

Subsequently, the infant was regularly controlled. At 18 months, nothing abnormal could be found, physical and mental development were fully appropriate.

Data for our further three cases (Nos 2, 4 and 5) are shown in Table I.

Thus, among the 5 infants with foetal hydrops, in one case no isoimmunization whatever could be demonstrated. Since the mother had a hydramnios and pregnancy toxaemia, the hydrops had to be ascribed to the hydramnios, as in the case reported earlier by DYGGVE [4]. In the other four patients, the hydrops was due to Rh isoimmunization.

In all the five cases, one or more artificial as well as several spontaneous abortions were mentioned in the history. None of the mothers had had two deliveries, and none had received anti-D IgG prophylaxis. All the infants were born before the 38th week of pregnancy, by Caesarean section. Patient M. I. (Case 5) had to be delivered in the 28th week in view of intrauterine damage. This was so grave that the baby could not be resuscitated. Another patient (No. 2) died at age 12 hours, after two exchange transfusions.

DISCUSSION

A few years ago, survival of a hydropic newborn was a rarity. OSKI AND NAIMAN in 1967 were still of the opinion that such patients can be kept alive for a few hours only. In the case of hydrops, the results of intrauterine transfusion are questionable and in any case much worse than if there is no hydrops. In LILEY's [7, 8] material of 13 hydropic patients who received intrauterine transfusions, none survived, whereas of 18 cases without hydrops, 13 survived. Recent results are, however, much more promising. According to GIRLING [6], despite the fact that of 56 newborns delivered subsequent to intrauterine transfusion, 42 were hydropic, nearly half of these could be saved by intensive treatment.

The main points of treatment, in agreement with the literature [2, 12], are as follows. The infant must immediately be intubated (when the oedematous larynx may present difficulties) and after clearing the airways by aspiration, intermittent positive pressure breathing has to be instituted. Then a catheter is passed into the umbilical vein, and to decrease portal pressure, blood is removed and its volume replaced by packed red cell mass. The next task is to remove the ascites fluid through a paracentesis in the left iliac fossa (on the right side one may injure the large liver). This intervention should be carried out even when an intrauterine transfusion has been performed and also when ascites cannot be detected by percussion.

TABLE I

Hydrops foetalis

Initials, Patient	Mother's blood group	Earlier pregnancies	Gestational week	Birth weight	Blood group	Course	Notes
1 M. Gy.	B Rh pos.	1 abortion 1 premature delivery	38	2750	B Rh pos.	Died at start of exchange transfusion	For details, see text
2 Sz. G.	A Rh neg.	2 abortions	30	1900	A Rh pos.	Severe Rh isoimmunization. Caesarean section. Hydrops. Extreme anaemia. Two exchange transfusions. Died after 36 hours	Low platelet count
3 L. L.	AB Rh neg.	1 stillborn 1 abortion	36	2000	B Rh pos.	Rh isoimmunization. Caesarean section. Poor condition. After 3 exchange transfusions and intensive care, survived.	Good condition healthy at 18 months Details in text
4 Cs. B.	AB Rh neg.	1 abortion	35	2300	B Rh pos.	Rh isoimmunization. Caesarean section. Oedema. Extreme anaemia. Two exchange transfusions. Survived.	Good condition at one and a half year
5 M. I.	A Rh neg.	1 abortion 1 twin and 1 preterm —stillbirth	28	1500	A Rh pos.	Rh isoimmunization. Caesarean section. Very poor condition. Hydrops. Resuscitation unsuccessful.	

When in the above way an equilibrium has been ensured and digitalization and furosemide treatment were started, an exchange transfusion is performed and eventually repeated according to the laboratory data. After the satisfactory control of hyperbilirubinaemia, an eventual anaemia, hypoglycaemia and hypoproteinaemia may need correction.

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