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Paper

The Emerging Pattern Of Hydrops Fetalis - Incidence, aetiology and management.

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ABSTRACT

***Objectives* – To analyse the incidence, aetiology and management of live born cases of hydrops fetalis in a Regional Perinatal Centre.**

***Methods* – We reviewed 35 cases of hydrops delivered over a six year period.**

***Results* – Non-immune hydrops accounted for 80% of the cases and the majority of babies required Level 1 intensive care. The mortality rate was 40%.**

***Conclusion* – The pattern of hydrops is changing. Most of these babies now have non-immune hydrops and approximately two thirds are surviving.**

KEY WORDS: Hydrops Fetalis, incidence, management

INTRODUCTION

Hydrops fetalis describes the fetus with generalised subcutaneous oedema and fluid collections in some or all serous cavities. Ballantyne described the first case of hydrops fetalis over 100 years ago and 50 years later Potter[1] described non-immune hydrops. Now over 80 conditions are known to be associated with hydrops.[2] Historically, Rhesus isoimmunisation was the leading cause of hydrops in the newborn. However, with the institution of passive maternal immunisation and the development of intrauterine fetal transfusions over the last few decades, non immune hydrops has become relatively more common.

The incidence, aetiology, management and outcome of hydropic babies born in the period 1974-1989 was published over 10 years ago.[3] Since then, obstetric and neonatal practices have changed. The aim of this study was to review all live born cases of fetal hydrops in Royal Maternity Hospital over a six-year period to assess the changes and compare current with previous mortality rates.

METHODS

We performed a retrospective hospital chart review of all live born cases of hydrops delivered in Royal Maternity Hospital, Belfast in the period 1996-2002. Stillbirths were not included because the cause had not been identified in some cases. The appropriate case notes were identified using both computerised ICD10 coding system and manual review of the admission logbook.

A proforma was then completed and the following information recorded – gestational age at diagnosis, delivery details, birth weight, aetiology (if known), and subsequent management and outcome.

RESULTS

In the six year period there were 35 live born cases of fetal hydrops among the 25,443 live born deliveries in Royal Maternity Hospital – an incidence of 1.34 /1000 live births. We chose not to include stillbirths because, in some cases, a full assessment to determine the cause had not been performed.

An antenatal diagnosis of hydrops fetalis was made by ultrasound scanning in 25 out of the 35 cases with the median (range) gestational age at detection being 26.8 (16-33) weeks. Out of the 10 cases that were not detected antenatally, one case was due to Rhesus disease and the remaining nine were in the non-immune group (one Trisomy 21 and eight unknown cause). Eight women had amnioreductions, one had an intrauterine fetal transfusion and one had both.

The median (range) gestational age at delivery in our study was 31.5 (26-38) weeks and the birth weight was 2371 (882-4844) grams. Male to female ratio was 2.5:1

Non-immune hydrops accounted for 80% of the total diagnoses. In four cases a cardiac cause was found (two supraventricular tachycardias, one dilated cardiomyopathy and one double outlet right ventricle and transposition). Four babies were found to have Trisomy 21. One baby was found

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to be CMV positive, one had a diaphragmatic hernia and one baby had a thoracic myofibroma. No cause was found in the other 17 cases.

The remaining 20% of cases were due to Rhesus incompatibility. None were due to other types of blood group incompatibilities.

The majority of babies were mechanically ventilated with 63% requiring chest aspiration or drains inserted early on in the course of their management. 13 babies (37%) had peritoneal aspiration performed or intraperitoneal drains inserted. Four of the seven immune babies had exchange transfusions carried out. Inotropes were necessary in 34% cases.

Over the six year period, the mortality rate for babies born with hydrops fetalis was 40%. There were 13 neonatal deaths (mostly occurring within 24 hrs of delivery) and one baby died at 62 days of age secondary to sepsis and renal failure. All 14 deaths occurred in babies in the non-immune group (three babies with Trisomy 21, one with thoracic myofibroma, one CMV infection and in nine cases no cause known).

DISCUSSION

In the earlier study in Royal Maternity Hospital from1974-1989, there were 27 live born babies with hydrops among the 52,177 live births, an incidence of 0.52/1000 live births.[3] Stillbirths were not included in this study. Wilson et al showed that then the commonest cause of hydrops was Rhesus isoimmunisation and mortality rates were initially as high as 100% and then fell to 50% between1978-1989. The specialised neonatal unit opened in the Royal Maternity Hospital in 1978.

Our study has shown that 80% of cases are now due to non-immune hydrops. New causes of non-immune hydrops are being reported every year but in approximately 50% of cases, no cause can be found despite extensive investigations. These babies continue to be born prematurely, though not as early as in the era when rhesus disease was a common problem. More babies receive ventilatory support and as the incidence of immune hydrops has decreased, there are fewer exchange transfusions. The mortality rate in our study had fallen to 40% (Table I).

The changing aetiology and improvement in outcome is due to several factors. The introduction of anti-D prophylaxis in 1969 has reduced the occurrence of Rhesus-D incompatibility but other materno-fetal blood group incompatibilities can occur.[4] The prenatal management of hydrops is improving with advances in fetal medicine such as the use of high resolution ultrasound scanning, fetal echocardiography and amniocentesis and cordocentesis to determine karyotypes. The information provided by detailed scans and investigations may direct the clinician towards appropriate fetal treatments such as intrauterine fetal transfusions if the fetus is anaemic or thoracocentesis if required. The decision to deliver the baby should involve close liaison between the obstetrician and he neonatologist. Planned delivery of the baby in a tertiary centre allows the baby the best chance for advanced resuscitation and neonatal care.[5, 6] This is reflected in the increased incidence rate we found in our study. That babies require extensive intensive care is evident in their need for ventilatory and inotropic support. The equipment available now is more advanced than the equipment that would have been available at the time of the previous study.[3]

A study carried out in a tertiary fetal medicine centre in Birmingham in the late 1990's also demonstrated that the majority of cases had a non-immune aetiology (87.3%). Mortality rates in this group were greater (62%)[7] than in our study.

The neurodevelopmental outcome of these babies is of concern. Recent studies have indicated a good outcome in survivors with non immune hydrops fetalis.[8] In a review of the long-term outcome in 19 children with non immune hydrops who survived beyond 1 year of age, 13 (68.4%) showed normal development, 2 mild developmental delay and the remaining 4 children had severe developmental problems.[9] We plan to further study the longterm outcome of survivors in our two cohorts of babies to determine whether their psychomotor development is comparable to that found by other researchers.

The authors have no conflict of interest

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TABLE I.

Comparisons of results with previous published findings

	1974-77[3]	1978-89[3]	1996-2002
Number	11	16	35
Gestation (wk) *	28.9±3.2	30.9±4.3	31.5±3.4
Birth weight (g) *	1607±741	1975±802	2371±795
Immune **	11 (100)	12 (75)	7 (20)
Non immune **	0	4 (25)	28 (80)
Mortality **	11 (100)	8 (50)	14 (40)

* mean ± sd ** N (%)