Short reports

Feto-fetal transfusion syndrome

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SUMMARY Out of 42 pairs of liveborn monochorial twins there were 32 pairs with vascular anastomoses. Of these, 11 pairs had feto-fetal transfusion syndrome. There were another 8 pairs of stillborn twin fetuses with vascular communications and in these chronic feto-fetal transfusion syndrome might have resulted in intrauterine death.

According to various authors vascular anastomosis is present in 85-100% of all monochorial placentae, and may allow the unbalanced transfer of blood from one twin to another.

Such vascular communications may be obvious on inspection or on injection of dye into the placental vessels. The most common anastomosis is of direct arterio-arterial type, but some are veno-venous. Perhaps of greater pathological significance is arterio-venous anastomosis between the two circulatory systems.

The feto-fetal transfusion syndrome (FFTS) is one of the contributory factors to the increased morbidity and mortality in monozygotic twin pregnancies. It has been suggested that two types of FFTS exist: a chronic form existing during pregnancy, and an acute form occurring only during parturition.

In chronic FFTS the donor twin, owing to an unbalanced transfer of blood, is generally hypervolaemic and anaemic, and shows varying degrees of growth retardation. In severe cases this twin may die in utero resulting in a fetus papyraceus at birth. The recipient twin however is hypervolaemic, polycythaemic, and is often the larger of the two. In severe cases this twin may develop cardiac hypertrophy and congestive cardiac failure. Furthermore increased urine production by this twin may lead to hydramnios and precipitate premature labour.

In acute FFTS the twins are generally similar in weight and length but one is polycythaemic and hypervolaemic and the other anaemic and hypovolaemic.

Patients and methods
During the period January 1975 to December 1980, 132 pairs of liveborn twins were admitted to the special care baby unit at the Royal Maternity Hospital, Glasgow. All had detailed pathological examination of their placenta and the presence of vascular communications was ascertained, if necessary, by the injection of Indian ink into the placental vessels. During the same period 24 pairs of stillborn fetuses were examined for the presence of vascular anastomosis. In liveborn monozygotic twins a difference of at least 5 g/dl between the haemoglobin levels of the two twins was taken as definitive evidence of FFTS. In stillborn twin fetuses the presence of pallor in one fetus and of redness in the other, without any other cause for such discordance, was considered as indicating the presence of FFTS.

Of the 132 pairs of liveborn twins studied, 42 pairs were found to have monochorial placenta and of these 32 pairs had vascular anastomoses (Table 1). In 21 out of these 32 pairs the pregnancy progressed as far as labour and delivery without any appreciable unbalancing of the fetal circulations and the twins were healthy at birth. In the remaining 11 liveborn twins varying degrees of FFTS occurred. The clinical and haematological details are shown in Table 2.

A difference of at least 5 g/dl between the haemoglobin levels of the twins was noted in every pair except in pairs 6 and 7. In these two pairs it was possible to check the haemoglobin level of only the donor twin. In Case 6 the recipient twin had died after 4 hours because of severe respiratory distress syndrome. In Case 7 the recipient twin had been healthy at birth and was not admitted to the special

<table>
<thead>
<tr>
<th>Type of anastomosis</th>
<th>No of placentae</th>
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<tbody>
<tr>
<td>Artero-arterial</td>
<td>20</td>
</tr>
<tr>
<td>Veno-venous</td>
<td>1</td>
</tr>
<tr>
<td>Artero-venous</td>
<td>2</td>
</tr>
<tr>
<td>Artero-arterial plus veno-venous</td>
<td>1</td>
</tr>
<tr>
<td>'Vascular links'</td>
<td>7</td>
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*Vascular links*= on injection dye passed from one part of the placenta to the other but the exact nature of the anastomosis was not ascertained.
care baby unit; unfortunately the haemoglobin concentration was not checked. The haemoglobin concentration of these two twins was low (Case 6 twin II 12 g/dl; Case 7 twin I 10 g/dl), and in the absence of any other finding to explain this or the presence of a pronounced colour difference between the two halves of the placenta the possibility of acute FFTS was considered to be likely.

Twin pairs 1–7 did not require any immediate treatment for anaemia or polycythaemia. However Cases 8 (donor twin I), 9 (recipient twin II), 10 (recipient twin I), and 11 (recipient twin I) required treatment. In Cases 9, 10, and 11 the recipient twin was markedly polycythaemic and symptomatic, and was therefore treated with a partial exchange transfusion using fresh frozen plasma. Of the donor twins Case 8 was shocked and pale at birth despite an initially normal haemoglobin concentration and packed cell volume (PCV) but showed a striking response to an urgent blood transfusion. In Case 6, donor twin II, the primary cause of death was severe respiratory distress syndrome although anaemic anoxia may have been contributory.

Of the 24 pairs of stillborn twin fetuses 8 had vascular communications. These are the examples of chronic FFTS resulting in intrauterine death in 6 pregnancies before 28 weeks’ gestation and in the remaining two at 35 weeks’ gestation. It is interesting to note that in three pairs polycythaemia occurred in the lighter twin. Also of note is that in four cases hydramnios was present during pregnancy.

### Discussion

This report demonstrates some of the features of both acute and chronic FFTS and compares these with similar reports from others. When pregnancy continues despite chronic FFTS the infants are likely to show pronounced differences in birthweight in addition to marked anaemia and polycythaemia. According to Tan et al., a weight difference greater than 20% is highly suggestive of the presence of FFTS. In our series, in Cases 2, 10, and 11 (Table 2) the weight of the smaller twin was less than 80% that of the large one, and as already noted, in two (Cases 10 and 11) the recipient twin needed a partial exchange transfusion because of severe polycythaemia.

The minimal weight difference between the twins in pairs 8 and 9 indicates that their blood supply was well balanced throughout pregnancy and that FFTS must have occurred during parturition. However, FFTS still resulted in severe polycythaemia in the recipient twin in pair 9. This twin required a partial exchange transfusion using fresh frozen plasma to reduce the PCV to about 60%. In pair 8 the donor twin was in a state of hypovolaemic shock at birth despite a normal haemoglobin concentration and PCV, and showed a striking response to an urgent blood transfusion. This suggests that blood loss had occurred just before delivery so that immediately after birth haemodilution had not taken place.

According to some authors it is rare for the donor twin to require immediate treatment because of severe anaemia. In our study all liveborn donor twins, except Case 8, tolerated a low haemoglobin level without requiring a blood transfusion. Probably in these cases FFTS had occurred slowly enough for compensation to occur.

More commonly it is the recipient twin who requires treatment because of polycythaemia. When the haematocrit is above 75% signs of hyperviscosity may occur, as happened in two of our cases (Cases 9 and 10). Some authors recommend that such cases should be treated whether they are symptomatic or not, on the basis that delayed intervention may be too late if irreversible changes are to be avoided. Oski and Naiman recommend using the following formula in working out the exact volume of blood to be exchanged for fresh frozen plasma:

\[
\text{Volume of exchange (ml)} = \text{blood volume} \times \left(\frac{\text{observed PCV} - \text{desired PCV}}{\text{observed PCV}}\right)
\]

In cases of chronic FFTS, if intrauterine death occurs a reversal of the shunting of blood may take place across the anastomosis. It is known that if the donor fetus dies, the live fetus maintains the circulation going through the dead fetus’ placenta, so that
the morphological changes of intrauterine death do not occur in this placenta. It may be that at about the time of death of the donor twin the cardiac output and blood pressure fall, resulting in the reversal of the shunting of blood, which then flows back from the polycythaemic infant to the donor.

In liveborn cases Klebe and Ingomar suggest that the second twin receives a larger placento-fetal transfusion than normal at the time of birth because blood from both parts of the placenta is transfused into the baby, particularly if there is a delay in clamping the cord. This would explain our findings in Cases 2 and 3 (Table 2). It seems that FFTS accounts partly for the greater fetal mortality and morbidity seen in monochorial as opposed to dichorial twin gestation.

We thank Dr J C MacLaurin for allowing us to study his patients and Mrs E Stewart for secretarial help.

Valvulitis—bacterial or rheumatic?

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SUMMARY An 11-year-old girl presented with pyrexia, severe mitral regurgitation, and cardiac failure. The child's condition deteriorated necessitating an emergency life-saving valve replacement. Although the revised Jones's criteria were not fulfilled, histology confirmed acute rheumatic carditis.

Most observers believe that acute rheumatic fever is on the decline in the West. In consequence the classical picture of acute rheumatic fever with carditis is now rare. Acute rheumatic heart disease none the less is not extinct and when it does occur it may be atypical in its clinical manifestations. We report here the case of an 11-year-old girl, mistakenly believed to have subacute bacterial endocarditis.

Case report

The patient, who had previously been well, presented with a 3-week history of lethargy, loss of appetite, and pharyngitis. There was some myalgia and vague complaints of joint pain, but there was no swelling or tenderness and at no time was a rash seen. One week before admission she developed a pyrexia of 39.4°C with rigors, and was treated with erythromycin and ampicillin. Despite this her condition continued to deteriorate with persistent pyrexia and the development of dyspnoea.

On admission she was pyrexial and dyspnoeic but no abnormality was found in the respiratory system. Examination of the cardiovascular system showed a sinus tachycardia, systolic thrill, and a pansystolic murmur maximal at the apex. There was no increase in the jugular venous pressure but the liver was enlarged 3 cm and was tender. The girl was pale but there was no lymphadenopathy or splenomegaly, and subcutaneous nodules could not be felt. There were no rashes or splinter haemorrhages. Joints were not swollen or tender, and the throat appeared normal. A working diagnosis of severe mitral regurgitation due to acute rheumatic fever, subacute bacterial endocarditis, or possibly a juvenile arthropathy with valvular involvement, was established.

Initial investigations revealed an erythrocyte sedimentation rate (ESR) of 150 mm in the 1st hour with a haemoglobin concentration of 9.8 g/dl and a white cell count of 11.1 x 10^9/l. Throat swabs and blood cultures were negative as was urine analysis. The autoantibody screen and RA latex tests were both negative. Antistreptolysin O titres on admission and 2 weeks later showed no increase. Chest x-ray film showed slight cardiomegaly but no pulmonary

References

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