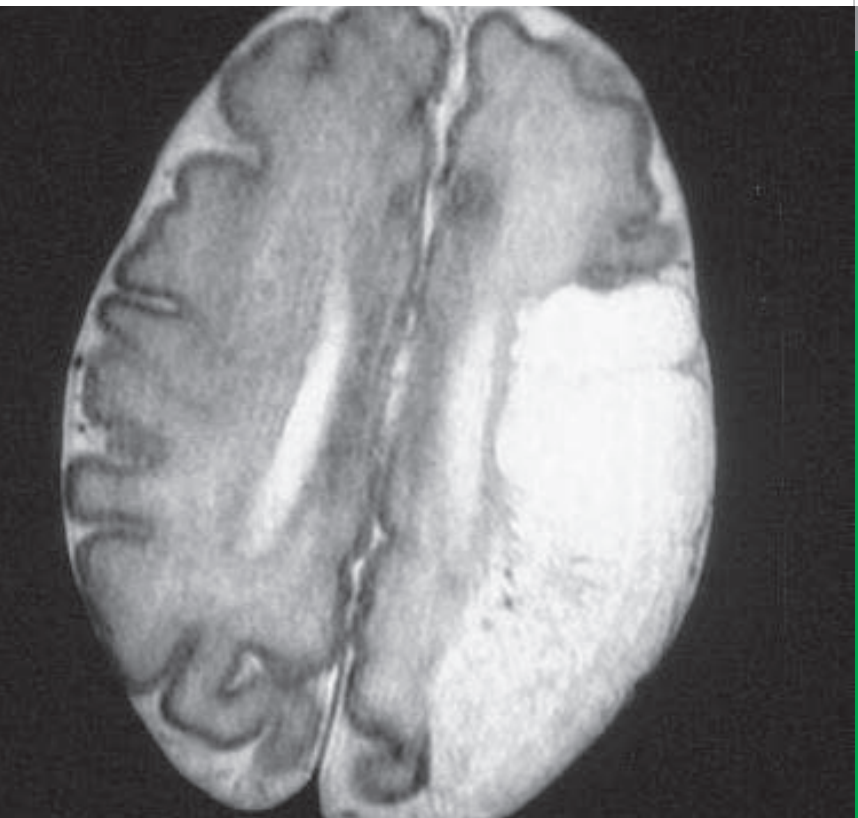
SWISS SOCIETY OF NEONATOLOGY

Surviving twin with encephalomalacia



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A 25-year-old G5/P1 (4 induced abortions in the past) became pregnant with twins. The current pregnancy was uneventful until 30 0/7 weeks of gestation when she developed acute abdominal pain. She consulted a gynecologist 4 days later when fetal death of one of the twins was noted. She was admitted to the University Hospital where polyhydramnios was diagnosed. Along with the fact that the surviving twin was much larger than the dead twin, a twin-to-twin transfusion syndrome was suspected. Decompression amniocentesis was performed and lung maturation induced with betamethasone. The mother went on to developed premature contractions and intravenous tocolysis was started. Doppler examination revealed a high maximal flow velocity in the middle cerebral artery as an indication for anemia in the surviving twin. At 30 6/7 weeks of gestation, a silent CTG with fetal tachycardia was an indication for emergency cesarean section.

Apgar score were 5 7, 8 and 9 at 1, 5 and 10 minutes, respectively. The male infant had a birthweight (1950 g), birth length and head circumference between the 75th and 90th percentiles. The dead twin had a weight of 1370 g (P 25). The liveborn twin was extremely pale and had generalized edema. Pulses and blood pressure were normal. He developed respiratory distress with a need for 40% oxygen which required treatment with nasal CPAP. Chest X-ray showed a small right-sided pleural effusion and normal cardiac size. In view of the benign course, the respiratory problem was interpreted

as wet lungs. A complete blood count revealed anemia with a hematocrit of 29% and evidence of a reactive bone marrow with reticulocytosis (15.2%, absolute count 383,000/µl) and increased erythroblasts (220/100 leucocytes). The baby was transfused within the first few hours of life with packed red blood cells. External signs of a disruption syndrome were absent. Abdominal sonography was normal. Cerebral ultrasound showed a sharply demarcated echogenic lesion in the left parieto-occipital region (Fig. 1). A cerebral infarct in the distribution area of the middle cerebral artery was suspected and confirmed with MRI (Fig. 2 and 3).

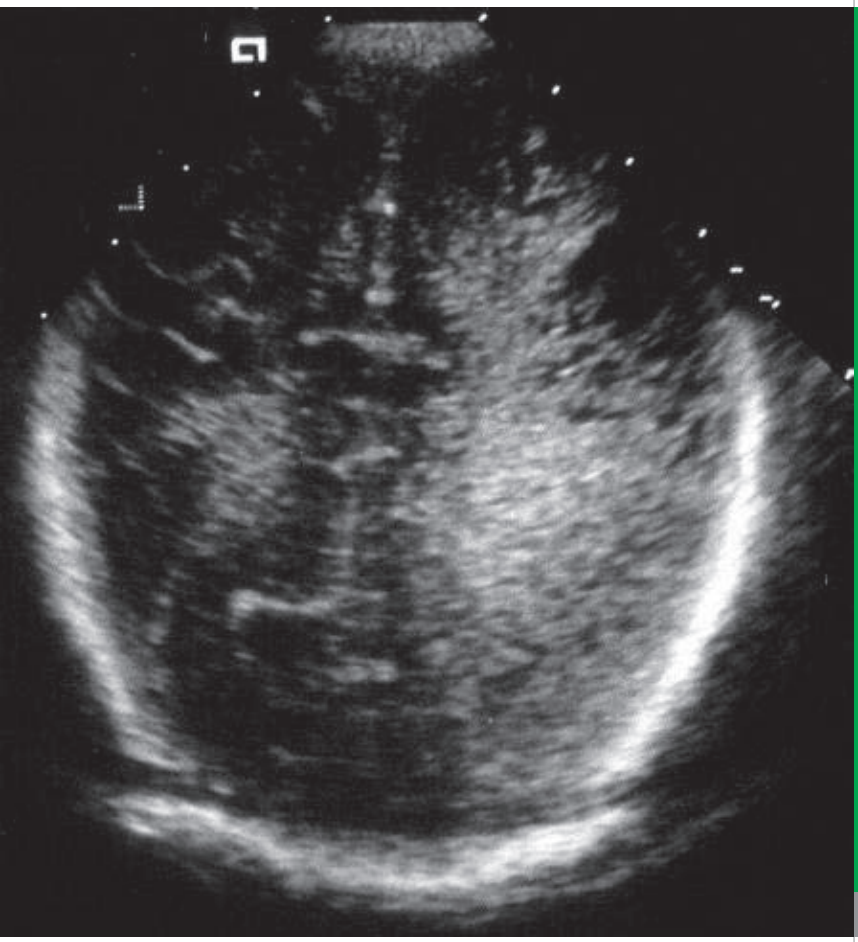

Fig. 1

US examination: hyperechogenic lesion in the left parieto-occipital region.

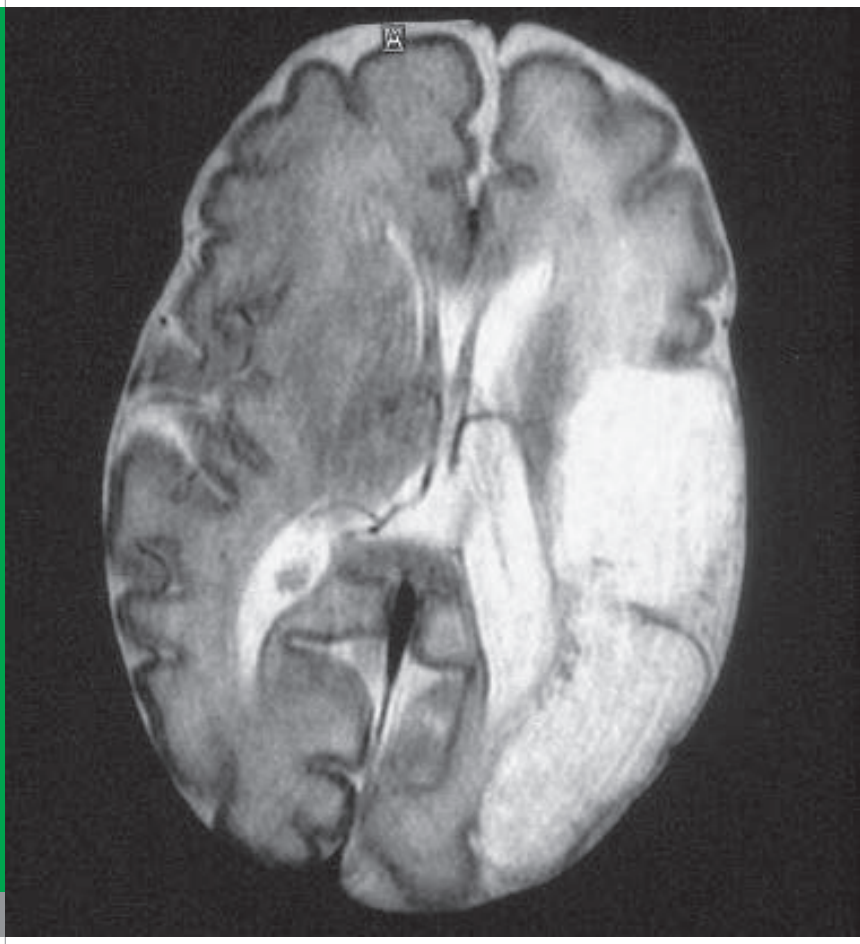

Fig. 2

MRI: T2-weighted images showing increased signal intensity in the left parieto-occipital region.

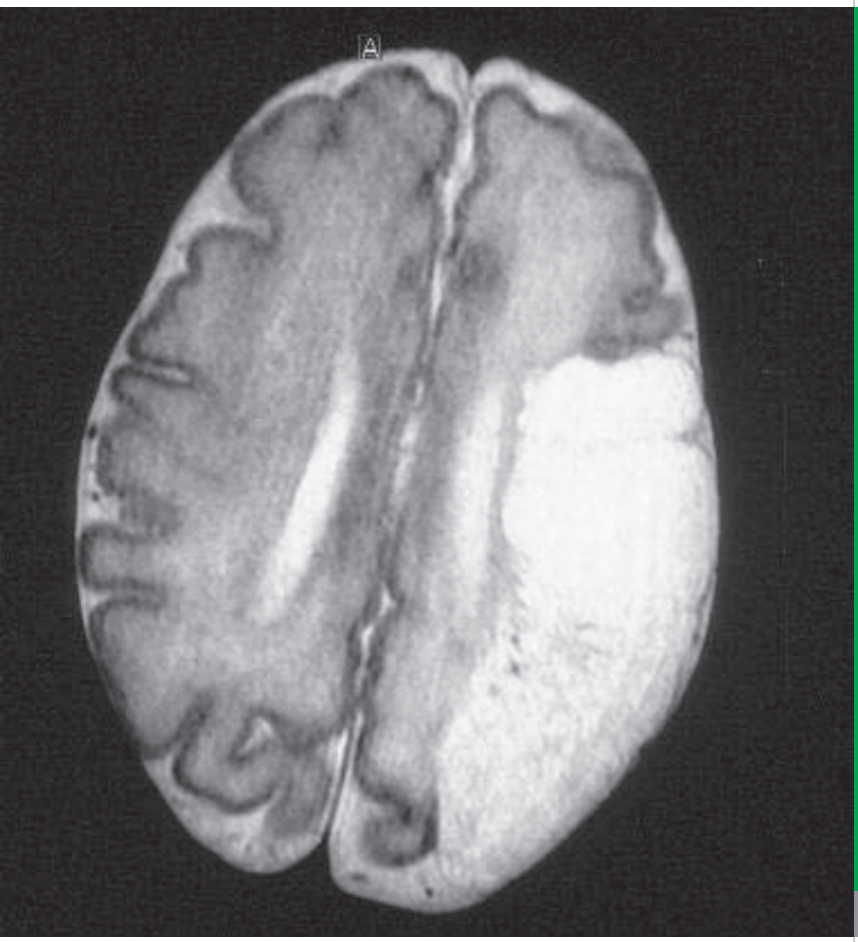

Fig. 3

MRI: T2-weighted images showing increased signal intensity in the left parieto-occipital region.

Neurological examination of the baby at term revealed increased extensor tone of the neck, trunk and lower extremities. A cerebral ultrasound examination at the same time revealed extensive, confluent multicystic lesions of the infarcted region. The lateral ventricles were not dilated.

DISCUSSION

Signs for twin-to-twin transfusion syndrome from the smaller (donor) to the larger twin (recipient) are polyhydramnios, a difference in birth weights, signs of hypervolemic (congestive) heart failure (generalized edema, pleural effusions) in the recipient twin. Placental examination reveals monochorial twins with the presence of blood vessel anastomoses.

The cause of fetal death in the smaller twin or donor is probably the result of increasing anemia and hypovolemia. However, at that point, a reversal of the shunt took place, that is, blood flowed from the survivor (the previous recipient) to the dead twin (the previous donor). As a result, the previous recipient developed severe anemia with hypovolemia and hypotension with hypoxic-ischemic tissue damage. The generalized edema could be explained by capillary leak through hypoxic damage of the endothelium. The severe hypotension probably led to the infarct in the middle cerebral artery.

Older theories about the causes of the so-called disruption syndrome are that thrombi from the dead twin pass through the blood vessel anastomoses and lead to infarcts in the various organs of the survivor.

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