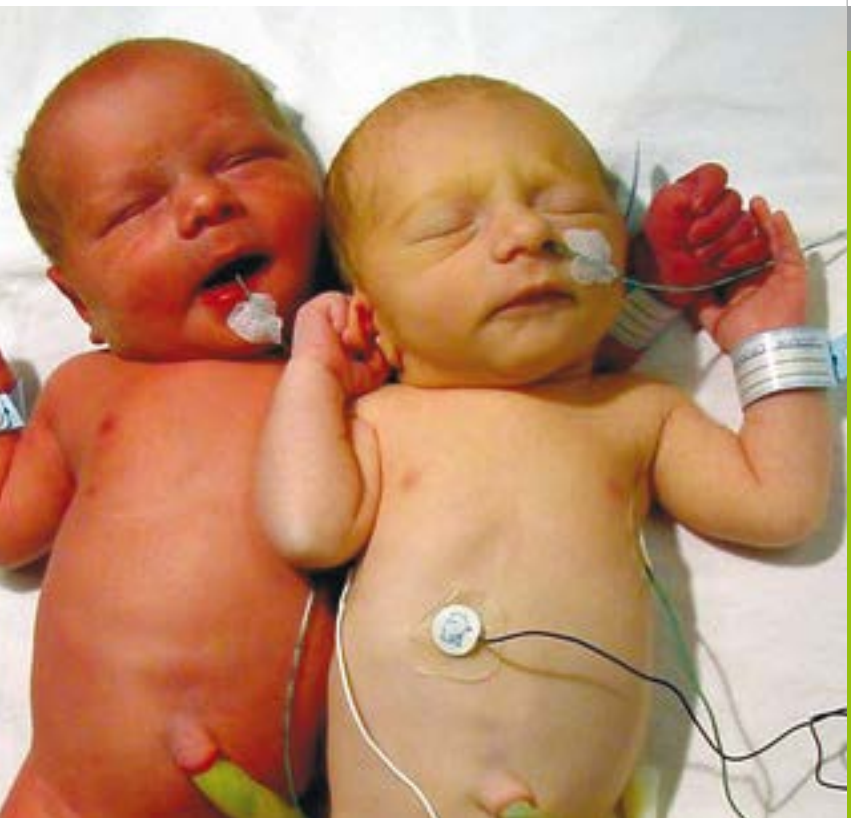
SWISS SOCIETY OF NEONATOLOGY

Twin-twin transfusion syndrome



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These female infants were born after an apparently uncomplicated monochorionic diamniotic twin pregnancy by primary caesarean section at 35 6/7 weeks at another institution. Postnatal adaptation was unremarkable for both twins (twin A: Apgar score 9, 10, 10; twin B: Apgar score 8, 9, 9 at 1, 5 and 10 minutes, respectively). Birth weights were 1850 g (P 3-10) and 2330 g (P 10-25) for twin A and B, respectively. After birth, an obvious difference in skin color was noted (Fig. 1-3) and capillary blood samples revealed anemia in twin A (hemoglobin 90 g/l, hematocrit 0.28) and polycythemia in twin B (hemoglobin 271 g/l, hematocrit 0.80). The infants were then referred to our institution for further management.

On admission, twin A was noted to be pale but hemodynamically compensated (heart rate 160 bpm, blood pressure 70/60 (mean 50) mmHg). She was breathing comfortably in room air with a rate of 50 breaths per minute. Her oxygen saturation was 94%. Anemia was confirmed with a venous hemoglobin of 70 g/l and a hematocrit of 0.21; in addition, there was a marked erythroblastosis (5.5 G/l, norm: 0-1 G/l). Twin B was noted to be plethoric but in no apparent distress with a respiratory rate of 45 breaths per minute and an oxygen saturation of 94% in room air. Her venous hemoglobin and hematocrit were 251 g/l and 0.71, respectively.

An obvious diagnosis of twin-twin transfusion syndrome was made. Cerebral ultrasound examinations were normal in both infants. The hemoglobin level of twin A was corrected by partial exchange transfusion with packed red blood cells (65 ml) to avoid hemodynamic compromise in a chronically anemic infant. In twin B, isovolemic hemodilution was performed using a partial exchange transfusion with normal saline (75 ml). The nurses noted an impressive difference of the blood stains on the newborn screening cards prior to the therapeutic interventions (Fig. 4). Both infants tolerated the procedures well and were discharged home at a corrected age of 37 4/7 weeks.

DISCUSSION

Before day 3 of pregnancy, twinning in monozygotic twins results in complete and hopefully equal separation of the chorion and eventually two separate placentas and gestational sacs (DCDA: dichorionic diamniotic twins). Embryonic division after day 3 results in a single shared placenta, known as monochorionic twinning. If the division occurs between day 3 and 7, there will be separate gestational sacs (MCDA: monochorionic diamniotic twins), After day 7, the fetuses will share a placenta and gestational sac (MCMA: monochorionic monoamniotic twins). If division occurs between day 13 and 15, it will be incomplete resulting in conjoined twins (1). Anastomoses are ubiquitous in monochorionic placentas; they can be deep arteriovenous, or superficial venovenous or arterioarterial connections. It

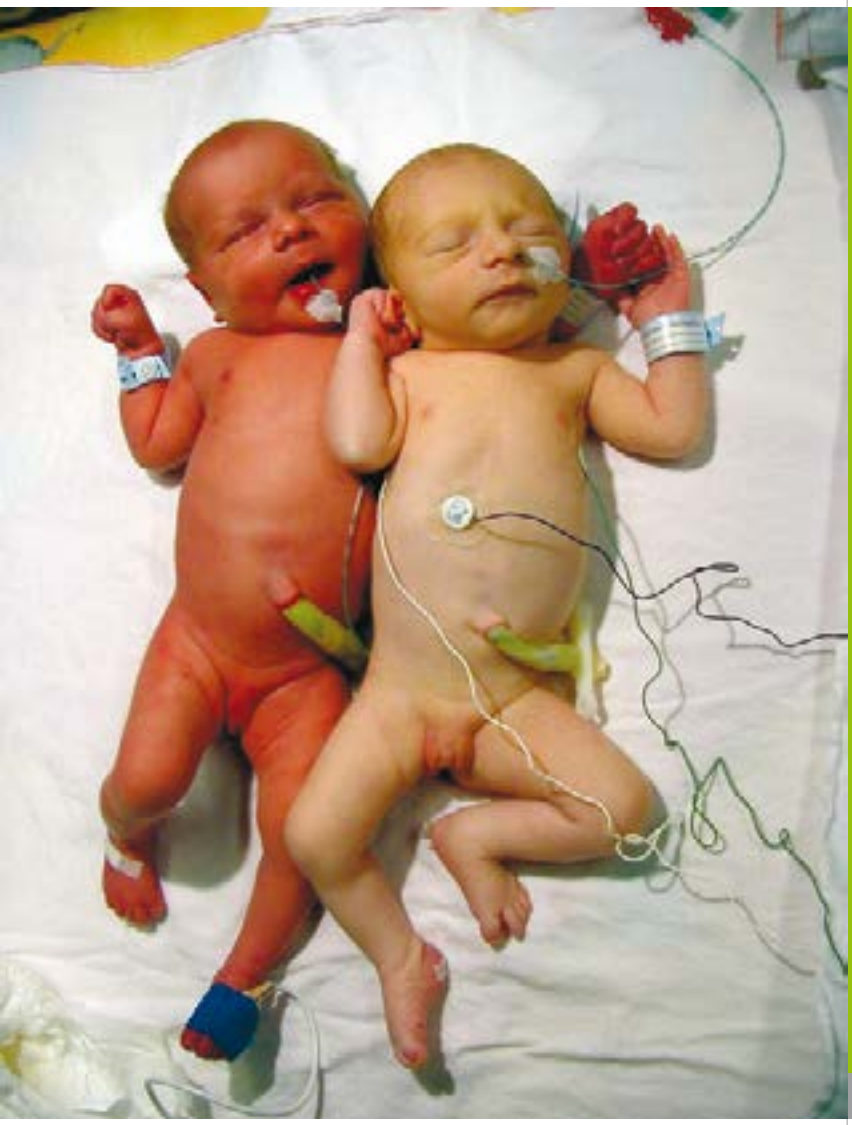


Fig. 1

Monochorionic twins at the age of 12 hours.

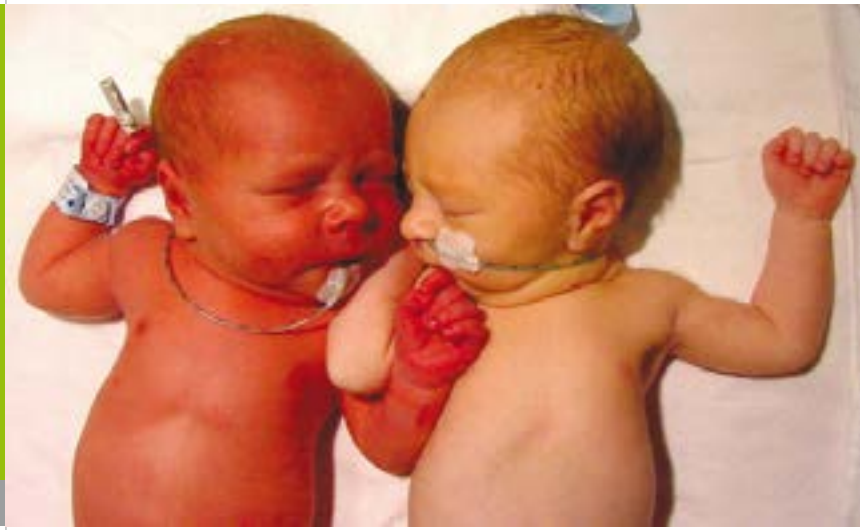
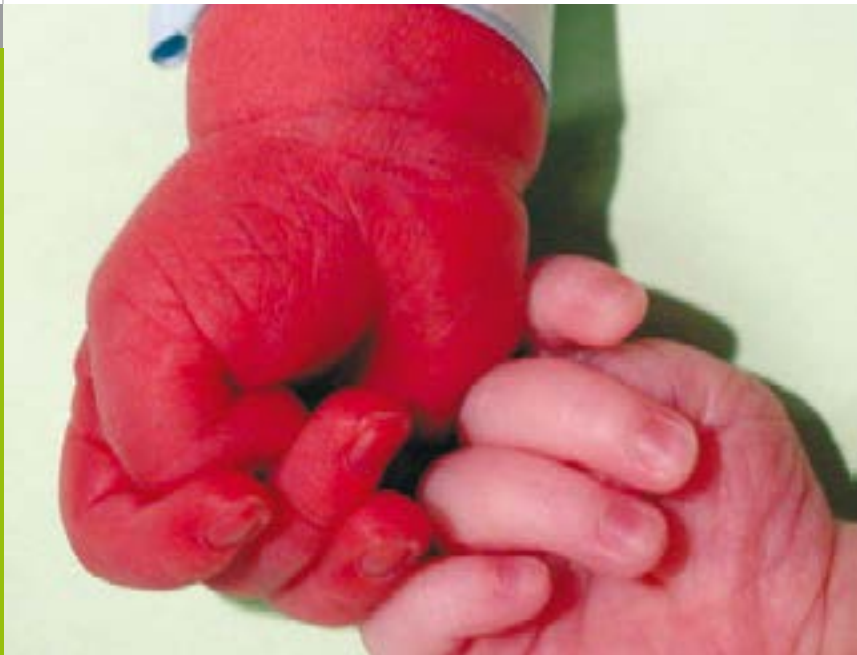


Fig. 2

Twin A (right, donor) and twin B (left, recipient) prior to exchange transfusions.

Obvious difference in skin color.

Fig. 3



is believed that unidirectional flow through arteriovenous anastomoses will eventually result in twin-twin transfusion syndrome unless there is compensation in the reverse direction (1). Twin-twin transfusion syndrome complicates approximately 15% of monochorionic twin pregnancies (2).

In the absence of intervention, twin-twin transfusion syndrome will result in vasoconstriction, oliguria, oligohydramnios, reduced growth rate, end-organ damage, long-term neurologic morbidity, and, often, fetal death in the donor fetus. The co-twin will experience volume overload, hydramnios, accelerated growth, cardiomegaly with cardiac decompensation, hydrops, and similar perinatal morbidity and mortality (1).

Pregnancies complicated by twin-twin transfusion syndrome should be referred to tertiary fetal medicine units experienced in the care of monochorionic pregnancies. In recent years, a range of new treatment modalities has been introduced into clinical practice, including amnioreduction, septostomy, selective reduction, and laser ablation (4). Amnioreduction offers good results in early stage disease (survival of one fetus 85%, survival of both fetuses 67%) (5). Laser treatment increases the proportion of single survivors, by reducing the number of both double survivors and double deaths (4).

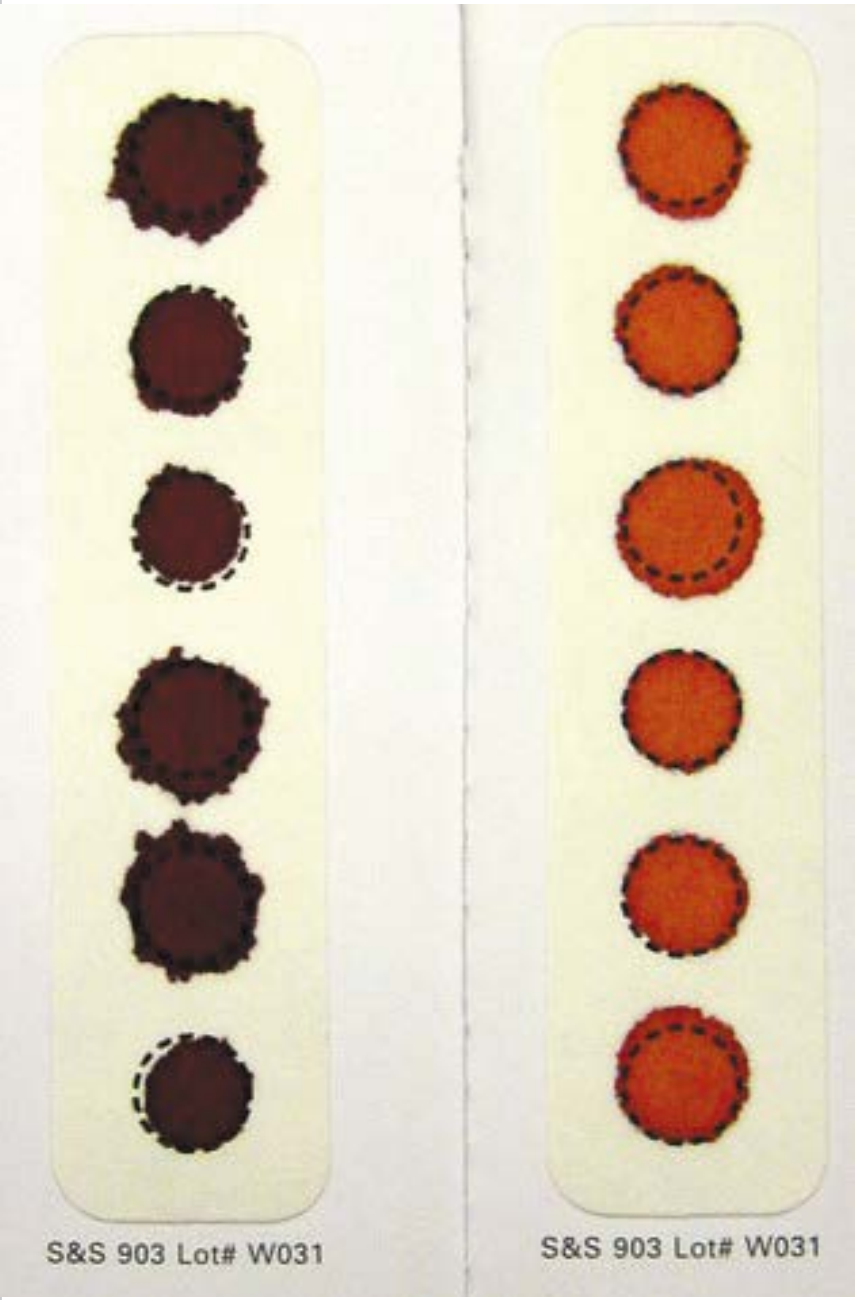


Fig. 4

Appearance of blood stains on Guthrie cards.

Postnatal management must focus on careful correction of anemia and polycythemia with partial exchange transfusion being the method of choice. Cardiac contractility can be impaired in both donor and recipient, and large fluctuations in intravascular volume should be avoided. Cerebral ultrasound examination and careful neurologic follow-up are mandatory for infants after twin-twin transfusion syndrome since there is a significantly increased risk of neurologic morbidity.

See also **Case of the Month 04/2004:** Unexpected severe anemia in an otherwise healthy newborn.

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