TWIN ANEMIA – POLYCYTHEMIA SEQUENCE (TAPS): POSTNATAL CORRECTION OF SEVERE ANEMIA IN THE DONOR TWIN BY TRANSFUSION OF BLOOD EVACUATED FROM THE RECIPIENT POLYCYTHEMIC TWIN

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ABSTRACT

Objective: To correct anemia by postnatal blood transfusion in the (intrauterine) donor twin neonate with blood obtained from the (intrauterine) recipient polycythemic twin.

Methods: The monochorionic twins displayed features of TAPS; concordant intrauterine normal amniotic sacs and fluid volumes, concordant birth weights (2.4kg and 2.2kg) but discordant postnatal haemoglobins (28.3d/dL and 7.3g/dL). Following resuscitation and admission to neonatal intensive care unit, clinical assessment revealed severe polycythemia and severe anemia in the first and second twin respectively. By partial exchange blood transfusion, blood obtained from the first twin was transfused to the second twin.

Results: The babies responded well to neonatal care and anemia was effectively corrected (Hb 18.0d/dL and 11.3g/dL for twin 1 and 2 respectively). One year after birth, the twins are doing well with appropriate developmental milestones.

Conclusions: Homologous transfusion of blood from a sister twin is an uncommon but welcome practice especially in a setting where donated blood is hardly readily available. This practice is strongly recommended in the management of neonatal anemia encountered in cases of TAPS and prenatal successfully managed Twin – Twin Transfusion Syndrome (TTTS).

Key words: Twin, Transfusion, Severe Anemia, Polycythemia

INTRODUCTION

The subject of multiplets, dichorionicity, monochorionicity (monoamniotic and diamniotic) (Fig. 1), is a major challenge to perinatologists because of the multiplicity of complications encountered, mostly attributable to placental vascular angioarchitectural pathology with resulting antenatal and perinatal complications and late sequelae. These complications include Twin - Twin Transfusion Syndrome (TTTS) or Twin Oligohydramnios Polyhydramnios Sequence (TOPS), Twin Anemia Polycythemia Sequence (TAPS), Conjoint Twins, and varied forms of twin monsters.

FIG 1

Twin Anemia-Polycythemia Sequence is a form of Twin-Twin Transfusion Syndrome (TTTS) in monochorionic twin gestation described by Lepriore and co-workers in 2007.[1] With reference to fetofetal transfusion syndrome in general, the pathology essentially consists of abnormal inter-twin placental fetal vascular anastomoses (Figs 2, 3), first described

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as 'Chorioangiopagus Vessels' by Friedrich Schatz in his study of monochorionic pregnancies between 1875 and 1910.[2] These chorioangiopagus vessels described the placental vascular anastomoses, which were considered variant on the nomenclature used for the description of conjoined twins. Schatz suggested that the twins were connected by blood vessels and this constituted the mildest form in the spectrum of what could be considered 'the Conjoint Twin Syndrome'. The increased morbidity and mortality rates in monochorionic twins are directly related to the angioarchitecture of the monochorionic placenta with its almost ever (95%) present vascular anastomoses.[3,4] With special reference to TAPS, it is characterized by randomly distributed vascular anastomoses of very tiny superficial blood vessels in the placenta complicated by inter twin transfusion, discordance of haemoglobin and red blood cell counts in the donor and recipient twins, and the absence of twin oligo/polyhydramnios (TOPS) discordance [1,5], a feature of TTTS.

FIGS 2 and 3

The actual incidence of the condition is not known for certain because of the limited knowledge of TAPS. Nonetheless, some institution-based surveys[,6,7,8] suggest that TAPS occurs spontaneously in 1%to 5% of cases of monochorionic pregnancies and in 1% to15% of all cases of TTTS treated by laser surgery (iatrogenic TAPS). TAPS is diagnosed in the antenatal period by the demonstration of abnormal Peak Systolic Velocities in the Middle Cerebral Arteries (MCA-PSV) of the donor and recipient twins respectively during Doppler Ultrasound Scan examination. Accordingly, MCA –PSV of 1.5MoM is synonymous with severe anemia in the donor twin, while a value < 1MoM is diagnostic of significant polycythemia in the recipient twin, in the absence of TOPS.[9]

Postnatal diagnosis of TAPS is based on the presence of chronic anemia and reticulocytosis in the donor twin as well as polycythemia in the recipient twin in addition to the demonstration of typical placental angioarchitectural abnormalities revealed by the injection of a colored dye.[7] Indeed, the hematological criteria for the postnatal diagnosis of TAPS is exemplified by inter twin haemoglobin difference of > 8gm/dL and Reticulocyte Count Ratio of 1.7. [10]

The outcomes of TAPS following various antenatal interventions are not clear because of relatively limited knowledge of the condition.[11] It is remarkable that previous reports on the therapy of TTTS (TOPS) and TAPS emphasized antenatal procedures including serial amniocentesis, photocoagulation of placental vascular anastomoses (feoscopic laser occlusion of chorioangiopagus vessels - FLOC)[12], amniotic septostomy, dichorionization, intrauterine transfusion[13], and selected feticide by cord occlusion, among others. This report describes our experience with a novel approach to the postnatal management of TAPS in a set of twins with virtually no weight discrepancy between the babies but with marked difference in the Packed Cell Volumes.

The management involved the correction of severe anemia in the donor twin with the blood evacuated from the recipient twin. This approach of postnatal treatment has not been described previously to the best of our knowledge. Moreover, it is being highlighted as an important life–saving postnatal procedure for TAPS especially in settings that lack efficient blood transfusion services, notably in developing countries. Likewise, this mode of management has potential to prevent infections such as Hepatitis, Malaria, HIV and Cytomegalovirus. Furthermore, it is proposed that this mode of treatment be utilized as part of several prenatal procedures in the management of TAPS.

CASE REPORTS

The babies were delivered vaginally at term to a 28 years old, and Para 4^{+0} mother in Rehoboth Medical Centre (RMH), Benin City, and transferred to Modic Medical Centre (MMC), Benin City, for continuation of management. The pregnancy was characterized by transient moderate hypertension early in the third trmester that was controlled with alfa-methyldopa, and mild primary postpartum haemorrhage that required no blood transfusion. There was no history of prolonged rupture of membranes or intrapartum pyrexia.

FIG 4

Twin I

Baby I, a female, was delivered by spontaneous vaginal delivery with severe birth asphyxia (APGAR scores 3 at 1 minute) and was actively resuscitated. On presentation in MMC, she appeared plethoric, Weight – 2.4kg, Length – 48.5cm; Occipito Frontal and Chest Circumferences – 34cm and 30cm respectively. She had grunting respiration and was dyspneic with flaring of alae nasi, intercostal and sub costal recessions. The respiratory rate was 64 cycles / minute; Pulse rate was 160 beats / minute. The lung fields were clinically clear; heart sounds were normal and had a grade 3 systolic murmur. The abdomen, extremities and genitalia were normal.

The Full Blood Count revealed PCV- 83%; Hb – 28.3g/dL; WBC 9.2 X 10^{9} / l with neutrophils 31%, lymphocytes 69%, and random blood sugar was 38mg/dl; blood group was O - Rhesus positive; chest radiograph showed bilateral pneumonic infiltrates. Cardiac echocardiography showed a structurally normal heart. A diagnosis of polycythemia complicated by neonatal respiratory distress was established.

The clinical conditions of the neonates, including the diagnoses (refer to Twin 11 below), were carefully explained to the parents and informed consents were obtained for planned further management. The treatment that entailed neonatal partial exchange blood transfusion for Twin 1 was explained to the couple and consent was obtained.

Twin 1 was given rapid infusion of 5ml / kg of 10% Dextrose in Water followed by a dilutional partial exchange blood transfusion aimed at reducing the hematocrit to 55% by replacing evacuated whole blood (30mls) with equivalent amount of 0.9% Normal saline using the following formula: [7,9]

Volume to be exchanged =
90 x Actual PCV – Expected PCV ÷ Actual PCV.
(90 refers to blood volume kg^{-1})

She responded favorably and satisfactorily to the procedures without any negative side effect or complication. Her condition remained stable and her post transfusion hematocrit values on the 2^{nd}, 3^{rd}, and 5^{th} days were 57%, 55% and 56% (Hb = 18g/dL) respectively.

Twin 11

Baby II, a female, was a breech delivery within ten minutes of the birth of Baby I. She was inactive at birth with moderate birth asphyxia (APGAR score of 4 at 1minute) and responded to active resuscitative measures. On presentation to MMC, she was pale and had petechial eruptions on the face and had shallow respirations. The weight was – 2.2kg, length – 49cm; and Occipito Frontal and Chest Circumferences – 33.5cm and 29cm respectively. The heart rate was 180 beats / minute; Respiratory rate – 72 cycles per minute. The lung fields revealed transmitted breath sounds but heart sounds and abdomen were normal. The anterior fontanel was full and not pulsating; Moro's reflex was sluggish. The genitalia revealed mature female external genitalia.

The Full Blood Count showed PCV- 21%; Hb – 7.3g/d/L; WBC-14.8 x 10^9/L; Neutrophils – 16%; Lymphocytes 84%, Platelet reduced on film, blood group – O-Rhesus positive; Random Blood Sugar – 40mg/dL. Chest radiograph, Trans-fontanel Ultrasound Scan and Echocardiogram revealed normal findings. A diagnosis of severe neonatal anemia complicated by cardio-respiratory failure was established.

The clinical condition of twin 11 with the diagnosis was carefully explained to the parents as was done with twin 1. Their informed consent for the planned correction of severe neonatal anemia was obtained.

The severely anemic Twin 11 was treated by the simultaneous transfusion of whole blood obtained from twin 1 together with on-going partial exchange blood transfusion (EBT). She tolerated the procedure very well with no untoward side effects.

The repeat FBC on second day post transfusion showed PCV 34%, Hb 11.3g/dL WBC 31.4x 10^9/l; Neutrophil - 34%, Lymphocyte - 66%, platelet reported as normal on film. The haematocrit values on the 3rd, 4th and 5th days were 33%; Hb 11.0g/dL and 33%; 11.0g/dL and 34%; 11.3g/dL respectively. The Baby was stable and vigorous at discharge on the 5th day of life with a Heart Rate of 130 beats / min and Respiratory Rate of 50 cycles / minute.

She remained stable, and made satisfactory progress and recovered fully from her cardio-respiratory decompensated state.

Follow up

The babies were reviewed regularly. At the age of one year, they had achieved all appropriate developmental milestones; they were walking efficiently and weighed 9.2Kg and 10Kg respectively. Figures 5 and 6 show recent pictures taken early September 2015.

FIGS. 5 and 6

DISCUSSION

The babies had similar weights and gender but markedly discordant haemoglobin values in keeping with TAPS. Accordingly, the Packed Cell Volumes of the donor and recipient babies were 21% (Hb - 6.8g/dL) and 83% (Hb - 27.6g/dL) respectively and this represented an inter twin difference > 20g/dl in consonance with Class 5, severe TAPS (Slaghekke, Kist et al, Mabuchi et al).[9] Consequently, the babies presented postnatally with marked pallor in the donor and significant plethora in the recipient twin leading to features of severe anemia and polycythemia respectively requiring immediate measures to avert untoward harmful outcomes.

Anemia in the donor twin was corrected with the blood that was evacuated from the recipient twin being treated simultaneously by partial exchange transfusion. It is noteworthy that the recipient twin provided the blood that was utilized to correct the anemia in the donor twin. Thus, utilizing the evacuated blood from the (in utero) recipient twin is tantamount to returning the blood "donated" by the (in utero) donor twin to her postnatally. The transfusions were tolerated satisfactorily by the babies and the clinical follow up of the twins have shown intact neurodevelopmental and cardiovascular functions.

Management of TAPS using intrauterine blood transfusion for the donor and partial exchange transfusion for the recipient is on record in literature.[12] This is probably the first description of this novel mode of postnatal blood transfusion treatment in the management of twin-twin transfusion in multiple pregnancies. This mode of treatment is our recommended first choice of donor blood in similar circumstances, especially, but not exclusively, in developing countries that lack efficient blood transfusion services. Besides, blood is not such a safe product these days, given the possibility of incomplete screening of donors' blood

and transmission of infections such as Hepatitis B, C viruses as well as Human Immunodeficiency Virus, Cytomegalovirus, and Malaria parasites.

RECOMMENDATION

Our experience in the management of these twins, though limited, provides a basis to recommend that blood be obtained from a recipient twin to correct anemia in the donor twin in the postnatal management of TAPS.

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Fig 1: Zygocity and Chorionicity; impact on fetal, perinatal, and neonatal outcome with late sequelae. (*Liesbeth Lewi and Jan Deprest*[3])

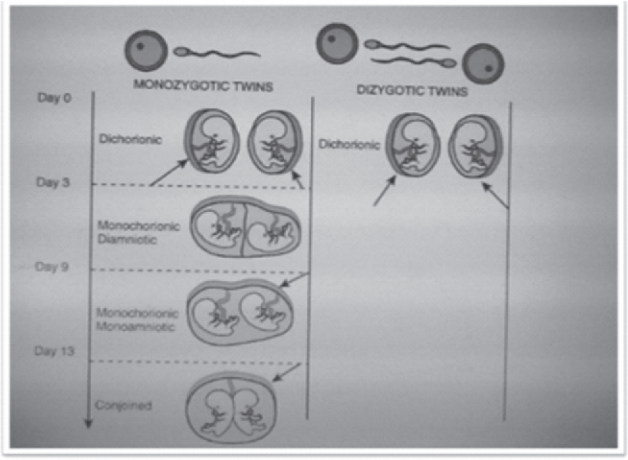

Fig 2: Monochorionic Monoamniotic Placental Angioarchitecture with Fetofetal Vascular Anastomoses; Macroscopic image of large diameter anastomoses and side-to-side insertion of umbilical cords. *(Liesbeth Lewi and Jan Deprest*[3]*).*

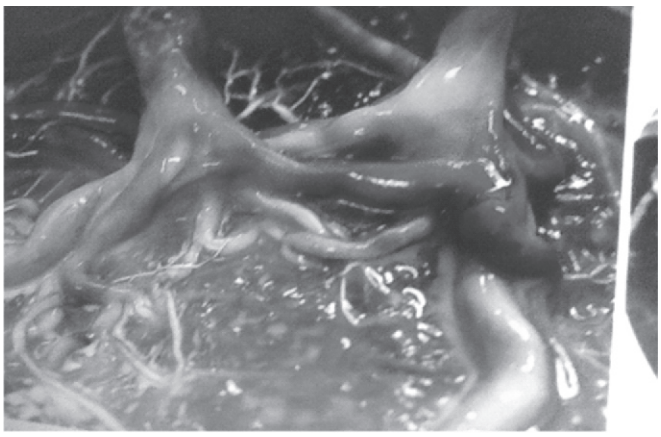

Fig 3: Angiography displaying Fetofetal Vascular Anastomoses. Diagnostic angiography in which injection of umbilical vessels of one twin was sufficient to visualize placental vascularization of both twins owing to presence of large arterioarterial and venovenous anastomoses. Also, about 50% of the placenta consisted of shared cotyledons. (*Liesbeth Lewi and Jan Deprest.*[3])

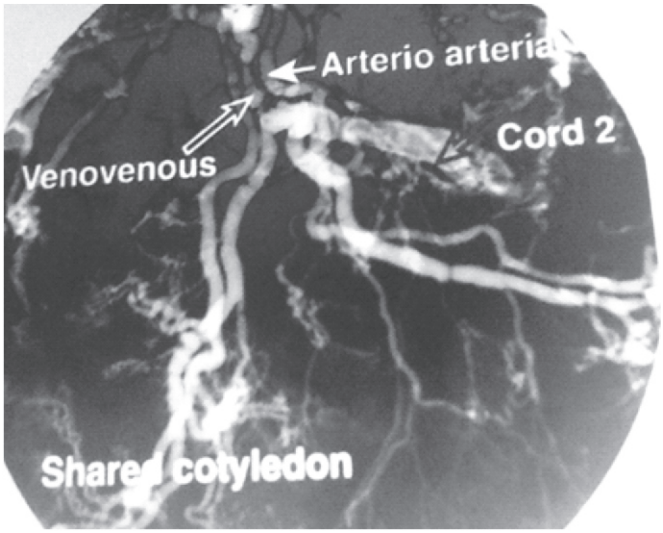

Fig 4: TAPS - Neonates at birth; Prenatal Donor (Left) and Recipient (Right) Twins.
(Osaghae DO and Unuigbe JA, 2015).

Fig 5: Twin 1 and 2 at the age 15 months

Fig 6: A happy family with the twins at the age of two years

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