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SUCCESSFUL MANAGEMENT OF A TWIN PREGNANCY COMPLICATED BY TRAP –A CASE REPORT

Gynaecology

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KEYWORDS

A 19-year-old woman visited our out patient department at 8 wks of amenorrhoea for a booking visit .She had conceived spontaneously Dating scan revealed a monochorionicdiamniotic twin pregnancy .Her past obstetric history was unremarkable. Her last child birth was 2yrsago.She had a spontaneous vaginal delivery at 38 wks of gestational age and delivered a healthy girl baby of 3.2kgs. Her menstrual history revealed that she had irregular cycles since menarche .At the booking visit she had no specific complaints .Her physical examination and investigations were within normal limits.Dating scan was done and the gestational age was reassigned (8 weeks+4 days) as she was unsure of her LMP .She was prescribed folic acid and asked to visit the clinic at 12 wks for an NT scan.

The patient had not turned up for an NT scan. She visited the clinic at 24wks of gestational age.Her ultrasound revealed a MONOCHORIONIC DIAMNIOTIC TWIN GESTATION with a viable fetus showing normal morphology and growth corresponding to gestational age.The placenta was located anteriorly .Foetal Doppler and Fetal echocardiography revealed normal parameters without any signs of cardiac failure (donor).TWIN A (Fig 1)

The second twin (recipient) TWIN B appeared grossly abnormal with hydropic features and no cardiac activity. This fetus had an incompletely formed skeleton, with no head, upper limbs, heart, or thoracic structures . It had an abdominal stump, with a cystic structure, possibly the urinary bladder, two abnormal lower limbs which showed massive, diffuse, soft tissue edema (Fig. 2).The supply for the perfused twin appeared to arise directly from the base of the cord insertion site of the pump twin .Doppler study revealed reversal of flow in the umbilical artery Ultrasound findings were consistent with TRAP sequence twin with an acardiusacephalus.

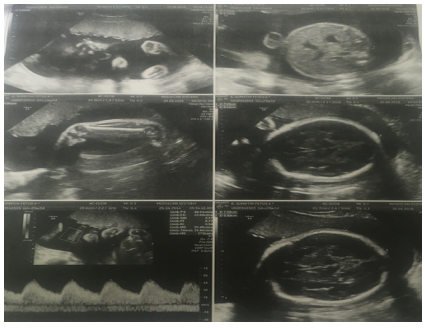


Fig 1- USG OF TWIN A SHOWING NORMAL MORPHOLOGY AND DOPPLER

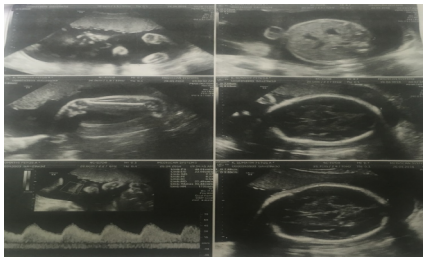


Fig 1

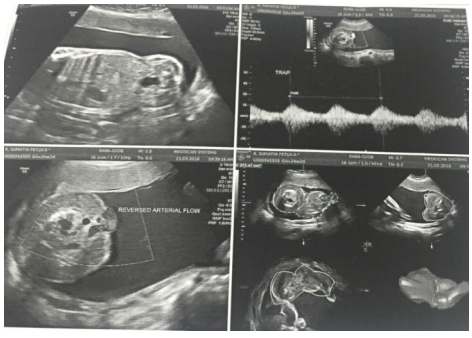


fig 2- USG OF TWIN B SHOWING GROSSLY ABNORMAL MORHOLOGY AND DOPPLER CHANGES

The scan findings and its implications were explained in detail to the family.They were explained about the treatment options of coagulation of the anastomotic vessels using an interstitial laser. Since the procedure ran a risk of miscarriage of about 10 %the patient and her family were apprehensive and were not willing to take any decision at that point of time.

A repeat Doppler done after 2 wks time showed an increase in liquor in Twin A with normal Doppler parameters .

She was under regular follow up with repeat ultrasound scans and doppler that were done fortnightly.The growth pattern of foetus A was within normal limits (fig 3).The pump twin did not demonstrate any complications of cardiac failure or hydrops. However the AFI of the pump twin showed a gradual increase



fig 2 –USG TWIN B SHOWIN G GROSS ABNORMAL MORPHOLOGY AND ABSENT CARDIAC ACTIVITY

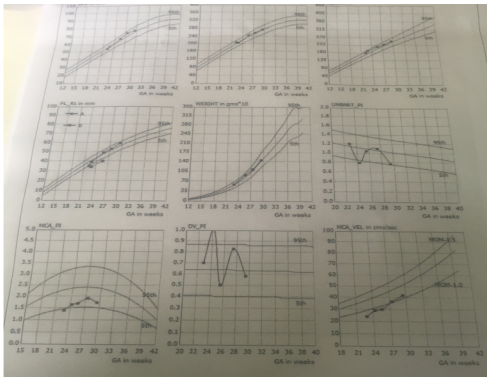


fig 3- GROWTH CURVES OF VARIOUS FOETAL PARAMETERS OF TWIN A

From a medical standpoint the gradually increasing polyhydramnios further heightened the chances of miscarriage/preterm labour /abruption if the procedure (laser coagulation)were to be attempted .After carefully analyzing the risk benefit ratio foetal medicine expert opined that a conservative approach would be a safer option provided the patient is kept under constant ultrasound surveillance with weekly Doppler and growth scans to monitor the growth of Twin A .

USG – 31 wks of gestational age
Twin A –Breech presentation
Polyhydramnios TWIN B -910 cc
SVP -11CM
Normal Doppler parameters
Efw- 1.8+-200 kg

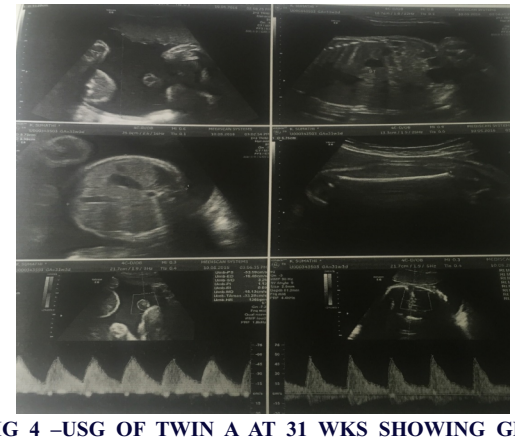


FIG 4 –USG OF TWIN A AT 31 WKS SHOWING GROSS POLYHYDRAMNIOS

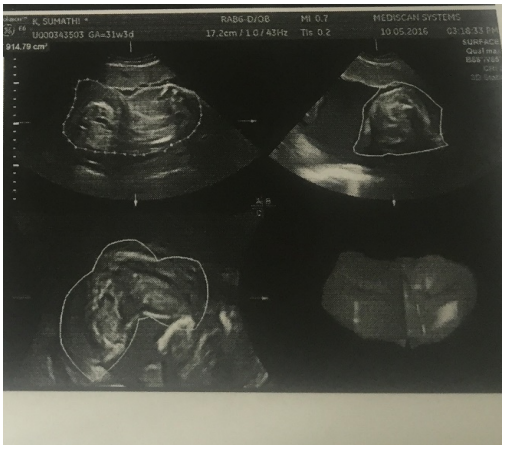


FIG 5 – USG OF TWIN B AT 31 WKS

At 34+4 weeks gestational age she had PPROM probably due to the gross polyhydramnios..She was started on Antibiotics and was given steroid cover to minimize the respiratory distress.In view of twin A being in Breech presentation she was delivered by a Caesarean Section .Twin A was an apparently normal live female baby weighing 2.2850 kgs with an Apgar score of 7/10 and 9/10 at 1 and 5 mis of birth respectively.

The acardiac twin appeared as a trunk like structure with birth weight 1100 gr. This baby had rudimental edematous lower limbs, pelvic bone, lower sacral vertebrae, and absence of thorax and cephalic structures (Fig 6)



fig 6 TWIN B

The placenta was unique and relatively large, was completely removed and there were no complications at birth. The female twin was discharged after 2 weeks in the intensive care unit because of neonatal respiratory distress syndrome. There was no neurological abnormality observed.The child is currently 2 yrs now and doing well with no health concerns.

DISCUSSION

Twin reversed arterial perfusion (TRAP) is a complication unique to monochorionic twins in which there is lack of a well-formed cardiac structure in one fetus (acardiac), abnormally perfused by a structurally normal co-twin (pump twin) through a superficial artery-to-artery placental anastomosis. Although the first case was described by Benedetti in 1533, very few cases have been reported in the literature since then (1)

The etiology and exact patho-physiological mechanisms are not well understood and the pathogenesis is controversial.

Two main hypotheses have been proposed (2):

1. Aberrant placental vasculature, with abnormal artery-to-artery placental anastomosis in early embryogenesis causes malformation of the acardiac twin (3). Imbalance in the blood pressure can lead to de-oxygenated blood-transfer from the healthy fetus (donor) to the impaired twin via the umbilical cord. This poorly oxygenated blood may maintain a certain growth of near-by located structures like the lower limbs or the intestines. In contrast, the blood-circuit in more distant areas like head, upper extremities, thorax is insufficient and their development often shows an arrest. So the receiving twin is gradually transformed into a parasite-like organism dependent on the blood supply from the donating twin(4).

2. A primary defect in embryogenesis in one twin leads first to failure of cardiac formation, followed by secondary development of anastomosis between umbilical vessels allowing survival of affected twin (5).

A range of structural anomalies have been reported in the acardiac twin, presumably due to the abnormal perfusion, from relatively normal body forms with well-differentiated organ structures through to apparent amorphous structures with no recognizable normal anatomy (6).

The heart may be completely absent (holoacardius), represented by a rudimentary cardiac structure (pseudoacardius) or less than 20% have identifiable cardiac tissue.

The antenatal diagnosis of TRAP-sequence during ultrasound is feasible and can be readily established during the first-trimester-screening (7).

An acardiac twin may be detected sonographically by showing fetal movement and growth, without definite differentiation of head and trunk. Commonly maldeveloped upper and lower extremities, without an evident heartbeat can be seen (8). The acardiac twin frequently has severe subcutaneous oedema and cystic hygromas, which can

significantly, increase the size of the fetus
Depending on the morphology of the acardiac fetus, four distinct types have been described in the literature (9):

1. acardiusacephalus: is the most common type, 60–75% of cases, with well-developed pelvis and lower limbs, but no head, usually no thoracic organs and often no arms

2. acardiusanceps: the most differentiated type of acardiac twins with well-developed body and extremities, but only a partially formed head and face; approximately 10% of cases

3. acardiusacormus: in which only cephalic structures were detectable with a close umbilical cord insertion; this is very rare, about 5% of cases

4. acardiusamorphus: in which the fetus is represented by a shapeless mass of tissue containing no recognizable structures; approximately 20% of cases.

Although this anatomical/pathological classification system has been widely used, it has no direct correlation with pregnancy outcome or implications for management, and it has been suggested that prenatal classification may be more appropriately based on the size of the acardiac twin and the apparent condition of the pump twin (9). Effectively the acardiac size may vary from incidental structures to large masses more than double the size of the pump twin, the largest reportedacardiac fetus weighing >6 kg

The overall perinatal mortality of pump twins is 50–55% and appears to correlate with the size of the acardiac twin. A higher weight of the recipient twin, with multiple internal organs, is more likely associated with a significant increase in the cardiac output and therefore perinatal mortality of the pump twin, generally due to cardiac insufficiency (10). This high cardiac output also increases perfusion of the fetal kidneys, resulting in overproduction of fetal urine and polyhydramnios, leading to premature delivery (before 32 weeks 'gestation) and prematurity (11).

The weight of the acardiac twin cannot be calculated using the standard formulae (such as Hadlock's), from the values of the head circumference, abdominal circumference, and femur length.

Instead, it is derived by the following formula: (1.2 × longest length 2) - (1.7 × longest length) (12).

Recently, Wong and Sepulveda suggested the role of abdominal circumference ratio as a prognostic factor in evaluating the impact of the acardiac fetus on the pump twin (10). When the ratio of the weight of the acardiac fetus to the weight of the donor fetus is greater than 70%, the incidence of preterm delivery is 90%, that of polyhydramnios is 40%, and that of congestive heart failure in the pump twin is 30%. In comparison, the corresponding rates are 75, 30, and 10%, respectively, when the ratio is less than 70% (10).

Doppler interrogation shows pathognomonic flow-pattern in terms of reversed arterial perfusion from the pumping twin towards the parasitic twin and in most of the cases (75%) reversed arterial blood supply enters the acardiac twin via a single umbilical artery. Venous blood-flow shows reversed perfusion as well (13)

Over the last two decades several procedures have been introduced aimed at establishing a permanent separation of the two blood-circuits of the twins improving prognosis and mortality in the pump fetus.

These techniques partly target the umbilical cord vessels using coil occlusion ligation, laser coagulation or bipolar coagulation. Other methods are aimed at the intrafetal umbilical vessels using alcohol injection, thermocoagulation, laser coagulation, radiofrequency or, very recently, high-intensity focused ultrasound (15). Laser coagulation of the vascular anastomoses on the placental surface has been proven as an effective and safe measure in many studies and case reports (16). Ultrasound-guided intrafetal needle approaches are easier and less invasive.

Nevertheless there is ongoing discussion regarding the correct management, expectant or not, and the exact intervention timing. TRAP sequence can be treated in 3 different ways, depending on the condition of the pumping twin (17)

- tailored approach with expectant management, until sonographic evidence of compromise in the pump twin (18) (polyhydramnios with a vertical pocket above 8 cm, cardiac dysfunction, abnormal Doppler-flow-patterns and/or hydrops); this expectant management has the advantage of avoiding an intervention that itself can cause miscarriage

- prophylactic intervention at 16–18 weeks after obliteration of the celomic cavity to reduce the risk of miscarriage (19)

- prophylactic intervention at 12 weeks to avoid the risk of death of the pump twin between 12 and 16 weeks

However the presence of relatively large weight acardiac twin, or monoamniotic twins are two parameters mandatory for early intervention Conservative management includes different tocolytic approaches and digitalis-treatment to support the cardiac performance of the pump twin (20). Regular sonographic evaluation of high-risk markers, including the amniotic fluid index of the pumping twin is recommended.

Amniotic drainages, or indomethacin-treatment may be performed to reduce the amniotic volume and to avoid premature labor, without notable side-effects , and improve prognosis in the survival fetus through reduction of the amount of amniotic fluid. The outcome of conservative management when acardius weight is < 50% of that of the pump twin, has been reported to be favorable in 88% of cases Some Authors, however, reported high pregnancy loss rates with the expectant management, even in cases of spontaneous cessation of flow in the acardiac twin. Authors reported loss rates, following diagnoses in the first trimester, of 83–100% with all losses occurring at ≤ 16 weeks The constantly increasing number of first-trimester diagnoses, together with the high loss rates reported in the pump twin if expectant management is chosen until 16 weeks, has raised the interest in earlier intervention in the first trimester and the recent studies add some evidence in favor of prophylactic intervention by intrafetal laser from 12 weeks onward.

Recently Berg et al. have registered in their experience fetal-loss rate from the first trimester onward was lower than previously reported. Expectant management in cases diagnosed at ≤ 14 weeks was associated with a fetal loss rate of 42.9%. After spontaneous cessation of flow to the TRAP twin, only 20.0% of pregnancies ended with death of the pump twin and there was no evidence of brain damage at the follow-up scans in the neonatal period In our case, the patient refused any operative procedures.Patient was under surveillance with weekly B-mode, and Color-Doppler examination.However a gross polyhydramnios later made the patient unfit for laser therapy and hence a conservative approach was resorted to What exactly causes progressive decompensation of a pump twin is unknown. Similar cases, treated with conservative management and without any developed complications, however, suggest a certain amount of resources to keep the blood-circuit of the pumping twin successfully working for quite a long time.

CONCLUSION

Monochorionic pregnancies should be carefully evaluated with first trimester ultrasound examination and TRAP sequence should be kept in mind as a complication of it. When TRAP sequence is diagnosed fetuses that are suitable for conservative management and fetuses which are candidates for interventional treatment should be selected carefully. Considerations should include fetal weight differences, amniotic fluid measure, and pump fetus well-being.

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