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Twin Anemia Polycythaemia Syndrome (TAPS) in a Monochorionic Twins with Dandy Walker Malformation in One Twin: A Rare Case Report

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Abstract
Twin anemia polycythemia sequence (TAPS) is a rare complication of monochorionic twins.in this, there is a net unbalanced intertwined blood transfusion in both the twins leading to anaemia in one twin and polycythemia in another twin. due to the result of this type of vascular disruption, sometimes brain malformations occur in one or both twins which can be life-threatening if not treated early in life. here is a case report of a twin pair with TAPS and Dandy walker malformation in one of the twins, for which an early successful ventriculoperitoneal shunt was applied.

INTRODUCTION

Monochorionic twin pregnancies are at increased risk for adverse outcome compared to dichorionic twin pregnancies and singletons. This is primarily due to the fact that almost all monochorionic twins share a single placenta, with inter-twin anastomoses allowing blood to flow bidirectionally between the two fetuses. Unbalanced net inter-twin blood transfusion may lead to various complications, including TTTS(twin- twin transfusion syndrome) and TAPS (twin anemia polycythemia sequence).[1]

TAPS is a newly described form of chronic and slow inter-twin blood transfusion characterized by large inter-twin Hb differences without signs of TOPS(Twin Oligo-Polyhydramnious Sequence).[1]TAPS may occur spontaneously (spontaneous TAPS) or after laser treatment for TTTS (post-laser TAPS). Spontaneous TAPS occurs in 3–5% of monochorionic twin pregnancies .[2] Whereas post-laser TAPS occurs in 2–16% of TTTS cases after incomplete laser treatment .[3] The wide range in incidence rate in post-laser TAPS can be explained by the use of different laser surgical techniques and/or the existence of different definitions and criteria for TAPS.

Twins have an increased risk for congenital malformations and disruptions, including defects in brain morphogenesis. These are separated into of malformations of cortical development (MCD), cerebellar malformations without MCD , and brain disruptions.[4] Cerebellar malformations without MCD varied from diffuse cerebellar hypoplasia to classic Dandy–Walker malformation. Recurrent association with twin–twin transfusion syndrome, twin anaemia polycythemia sequence (TAPS) intrauterine growth retardation, and other prenatal factors (chromosomal defects that affect fetal brain development,certain viral infections in the mother that pass to the developing baby,Exposure of the unborn baby to certain toxins or medications, Maternal diabetes) support disruption of vascular perfusion as the most likely unifying cause.[5] Here is a case of monochorionic diamniotic twins with TAPS and second twin having Dandy Walker Malformation , was operated at weight below 1800 gm with successful outcome.

CASE REPORT:

A 32 week twins born to a 24 year G2 P1 L1 mother ,spontaneous conception vaginal delivery. Antenatal scans were not available. Both are male babies with blood group B positive .Single placenta noted .Both the babies

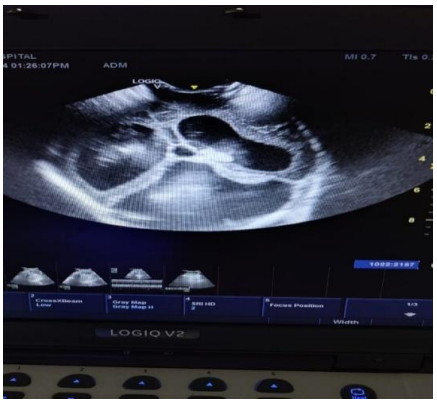
have respiratory distress grade II, required CPAP support after birth for 7 days. Both have late onset culture negative sepsis & required 10 days of antibiotics course.

Twin 1 [fig 1]with birth weight 1380 gm, have severe pallor with Hb = 7.2 gm%, Reticulocyte count-1.6%, peripheral blood smear suggestive of moderate microcytic hypochromic anaemia. This twin required 2 PRBC transfusion and iron supplements after that. This twin has also a large asymptomatic ASD of 7.1 mm .

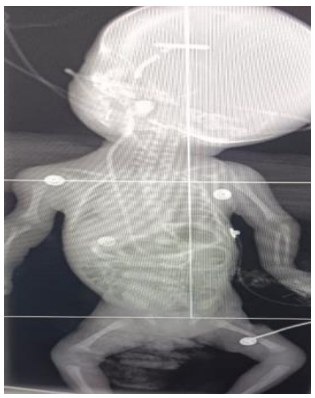
[Fig 1]

Twin 2 [fig 1]with birth weight 1480 gm, was plethoric with Hb = 18.5gm%, Reticulocyte count =3.5%, peripheral smear suggestive of mild poikilocytosis , normochromia & anisocytosis. Baby had HS-PDA (Hemodynamically significant PDA) of 2.3 mm, for which oral Ibuprofen was given followed by closure of the duct noted. By day 10 ,this twin has rapid progressive increase in head circumference .Anterior fontanelle- 5x5 cms,Posterior fontanelle-8x7 cm,metopic suture 1cm apart, coronal suture 0.5 cm apart,& lambdoid suture 1.5 cms apart.Head circumference =38 cm.Transillumination test-Positive[fig 2].

[Figure 2]



[figure 3]



[figure 4]

POCUS transcranial suggestive of Dandy Walker Malformation with moderate hydrocephalous[fig 3].MRI brain confirmed the USG finding.Hence neurosurgery opinion was sought.Even though this baby was less than 1800 gram ,in view of such congenital malformation which has always a threat of severe hydrocephalous,decision for ventriculo-peritoneal shunt taken [fig 4]. Post operative baby had aspiration pneumonia requirement of CPAP support for 5 days.head circumference comedown to 34 cm.

Twin KMC , A novel practice

[Figure 6]

Both the babies was on full feeds 4 days after admission, remained euglycemic throughout the hospital stay & was on twin KMC [fig 6]. Both were growing well on 1 month follow up. No shunt blockage or infection noted in twin 2. Head circumference remained static on follow up.

DISCUSSION:

Monochorionic twins share a single placenta and have many complications due to connections between placental blood vessels through various vascular anastomoses. Unbalanced inter-twin blood transfusion may lead to various complications, including twin-to-twin transfusion syndrome (TTTS) and twin anemia polycythemia sequence (TAPS). TAPS was first described less than a decade ago, and the pathogenesis of TAPS results from slow blood transfusion from donor to recipient through a few minuscule vascular anastomoses. This gradually leads to anemia in the donor and polycythemia in the recipient, in the absence of twin oligo-polyhydramnios sequence (TOPS). TAPS may occur spontaneously in 3–5% of monochorionic twins or after laser surgery for TTTS. The prevalence of post-laser TAPS varies from 2% to 16% of TTTS cases, depending on the rate of residual anastomoses.[1]

TABLE 1: Postnatal criteria for TAPS [7]

Postnatal stage	Intertwin Hb difference, g/dl
Stage 1	>8.0
Stage 2	>11.0
Stage 3	>14.0
Stage 4	>17.0
Stage 5	>20.0

TABLE 2 : Antenatal & postnatal criteria for TAPS [7]

Antenatal criteria	Postnatal criteria
MCA-PSV >1.5 MoM in the donor *and* MCA-PSV <1.0 MoM in the recipient	Intertwin Hb difference >8.0 g/dl *and* at least one of the following: - Reticulocyte count ratio >1.7 - Placenta with only small (diameter <1 mm) vascular anastomoses

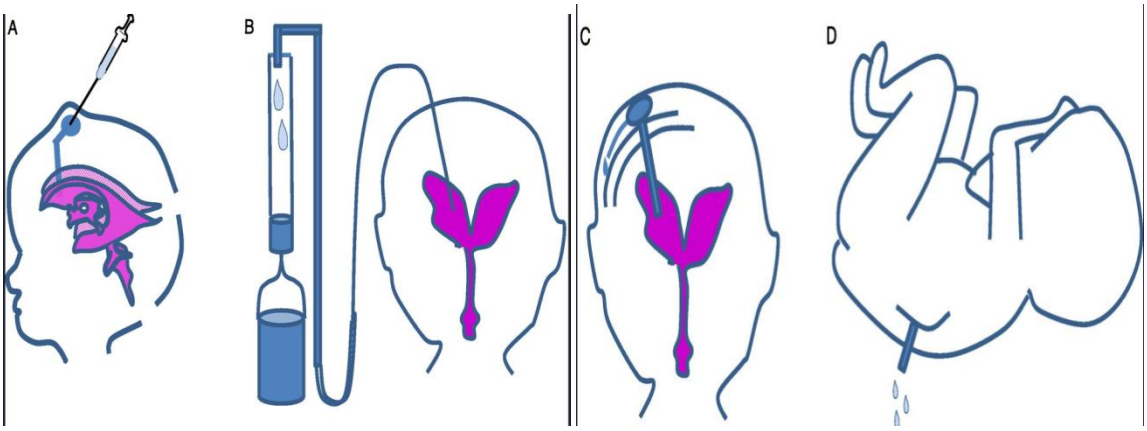
Thus our case falls into stage 2 TAPS, with Hb difference of 11.3gm% with Reticulocyte count ratio 2.18. We have managed with 2 PRBC transfusions for anaemic twin . For the polycythemic twin haematocrit was 60%. This does not required any partial exchange but 20 ml/kg additional fluid .There was hyperbilirubinaemia in polycythemic twin ,however managed with phototherapy over 3 days.

AAP recommendations (2014) for Pediatrics hydrocephalous management in preterm infants :

There are 7 recommendations [6]

RECOMMENDATION CONCERNING SURGICAL TEMPORIZING MEASURES: I. Ventricular access devices (VADs), external ventricular drains (EVDs), ventriculo-subgaleal (VSG) shunts, or lumbar punctures (LPs) are treatment options in the management of PHH. Clinical judgment is required. STRENGTH OF RECOMMENDATION: Level II, moderate degree of clinical certainty.[fig 7]
RECOMMENDATION CONCERNING SURGICAL TEMPORIZING MEASURES: II. The evidence demonstrates that VSG shunts reduce the need for daily CSF aspiration compared with VADs.

STRENGTH OF RECOMMENDATION: Level II, moderate degree of clinical certainty.
RECOMMENDATION CONCERNING ROUTINE USE OF SERIAL LUMBAR PUNCTURE: The routine use of serial lumbar puncture is not recommended to reduce the need for shunt placement or to avoid the progression of hydrocephalus in premature infants. STRENGTH OF RECOMMENDATION: Level I, high clinical certainty.
RECOMMENDATION CONCERNING NONSURGICAL TEMPORIZING AGENTS: I. Intraventricular thrombolytic agents including tissue plasminogen activator (tPA), urokinase, or streptokinase are not recommended as methods to reduce the need for shunt placement in premature infants with PHH. STRENGTH OF RECOMMENDATION: Level I, high clinical certainty.
RECOMMENDATION CONCERNING NONSURGICAL TEMPORIZING AGENTS. II. Acetazolamide and furosemide are not recommended as methods to reduce the need for shunt placement in premature infants with PHH. STRENGTH OF RECOMMENDATION: Level I, high clinical certainty.
RECOMMENDATION CONCERNING TIMING OF SHUNT PLACEMENT: There is insufficient evidence to recommend a specific weight or CSF parameter to direct the timing of shunt placement in premature infants with PHH. Clinical judgment is required. STRENGTH OF RECOMMENDATION: Level III, unclear clinical certainty.
RECOMMENDATION CONCERNING ENDOSCOPIC THIRD VENTRICULOSTOMY: There is insufficient evidence to recommend the use of endoscopic third ventriculostomy (ETV) in premature infants with posthemorrhagic hydrocephalus. STRENGTH OF RECOMMENDATION: Level III, unclear clinical certainty.



[Figure 7]

De vries et al [8] mentioned in his study that very early intervention prior to development of severe dialation of lateral ventricles have better neurological outcome. For VP shunt though exact timing is controversial, most of the units follows this procedure once infant reaches 2-2.5 kg. For <2 kg ventriculosubgaleal shunt or other temporising measures are preffered & permanent shunts after baby gains >2 kg.

As our case is a moderate hydrocephalous however associated with congenital brain malformation(DWM),permanent shunt was planned . Baby was 1700 gm when this procedure carried out . Intra-operative blood loss was minimum. Baby extubated immedietly after post op succesfully. Baby had requirement of low flow oxygen therapy for 7 days post op because of pneumonia . Even after 2 month followup baby is stable and growing adequately.In conclusion,for twins, especially monochorionic must be evaluated for TAPS (twin

anaemia polycythemia sequence)once clinical suspicion is raised. Also there should be high index of suspicion for congenital malformation of vital organs like heart ,brain and kidneys. Early permanent intervention of those congenital malformations are vital for better neuro- developmental outcome.

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