Twin-to-twin transfusion syndrome: a case report

Presenter: Dr. Kanika Sharma

- No conflict of interest

Birth Details

- Twin 1 (MCDA twins)
- Extreme preterm (27 weeks)
- ELBW baby (730 grams)- between 10^{th} and 50^{th} centiles by Fenton growth chart (expected fetal weight 630 grams without edema)
- Male baby
- Recipient twin/IUGR/Severe RDS/CHF
- Delivered via Emergency LSCS (Severe FGR with fetal heart decelerations with reversal of flow in ductus venosus) on 11/10/2020.

Antenatal history

- High risk pregnancy
- Advanced maternal age- 37 years
- Obesity
- ML x 10 years
- G3L1A1 mother with twin pregnancy (MCDA twins)
- IVF conception
- Referred case
- GDM (on insulin)
- Hypothyroidism
- First diagnosed to have TTS in recipient twin at 19 weeks of gestation
- Received a single dose of steroid 6 hours prior to delivery
- Amnioreduction done 8 hours prior to delivery

Course/progression of RDS

- Severely depressed at birth- HR 30/min, no spontaneous respiration
- Intubated in delivery room
- Shifted to NICU in a transport incubator
- Ventilatory settings: Mode SIMV- FiO2 100%, PIP 24, PEEP 6, VR 45
- Surfactant given
- All procedures including surfactant administration, UVC insertion, antibiotics and inotrope support initiation were completed in the first GOLDEN hour of life.

Investigations

Labs

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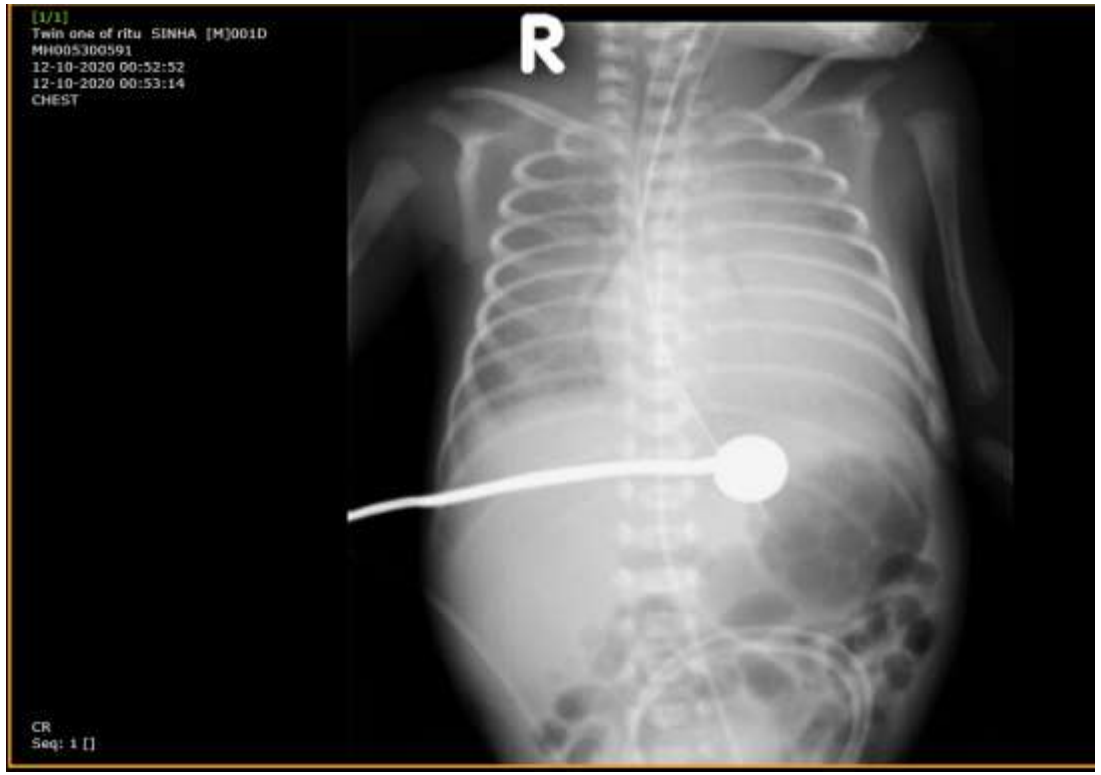
Chest X ray



- Inj. Dopamine and Dobutamine started at 5 mcg/kg/min through central access (poor perfusion, prolonged CRT, BP not recordable)
- Bedside ECHO showed poor cardiac contractility (biventricular dysfunction)
- Repeat surfactant given at 16 HOL (High ventilatory requirements FiO2 40% and PIP 20)

Management of CHF

- IVF started at 80 mL/kg/day
- Inj Dopamine stopped on DOL-3 but Dobutamine continued for poor cardiac contractility (gradually tapered and stopped on DOL-6)
- Repeat ECHO (DOL-6): Significantly improved cardiac contractility with LVEF 51-54%. PDA closed spontaneously.
- Polyuria UO 5-6 mL/kg/hour (adequate volume replacement done): twice daily baby weight monitoring and serum Na monitoring
- Careful fluid restriction in babies with PDA

- Extubation done on DOL-4 (SIMV settings: FiO2 30, PIP 16)
- Put on nasal CPAP support- FiO2 30%, PEEP 6 (target SpO2 91-95%)

Other problems

- Apnea of prematurity: Inj. Caffeine loading dose given within first 6 hours of life and maintenance dose continued
- Neonatal jaundice
- Mild BPD
- Stage 2 ROP

Nutrition

- OG feeding initiated from DOL-2 (EBM)
- Full feeds reached by DOL-8
- Fortification with HMF sachets (Pre Nan) started at DOL-6
- Reached birth weight by DOL-23
- Vaccination with BCG, Hep B at 34 weeks of gestation
- Discharge weight- 2.18 kg (37 weeks gestation)

- US cranium done according to protocol- normal study
- ROP screening Stage 2 ROP BE which regressed on follow up.
- Discharged at 37 weeks (DOL 89)

Twin 2 (donor twin)

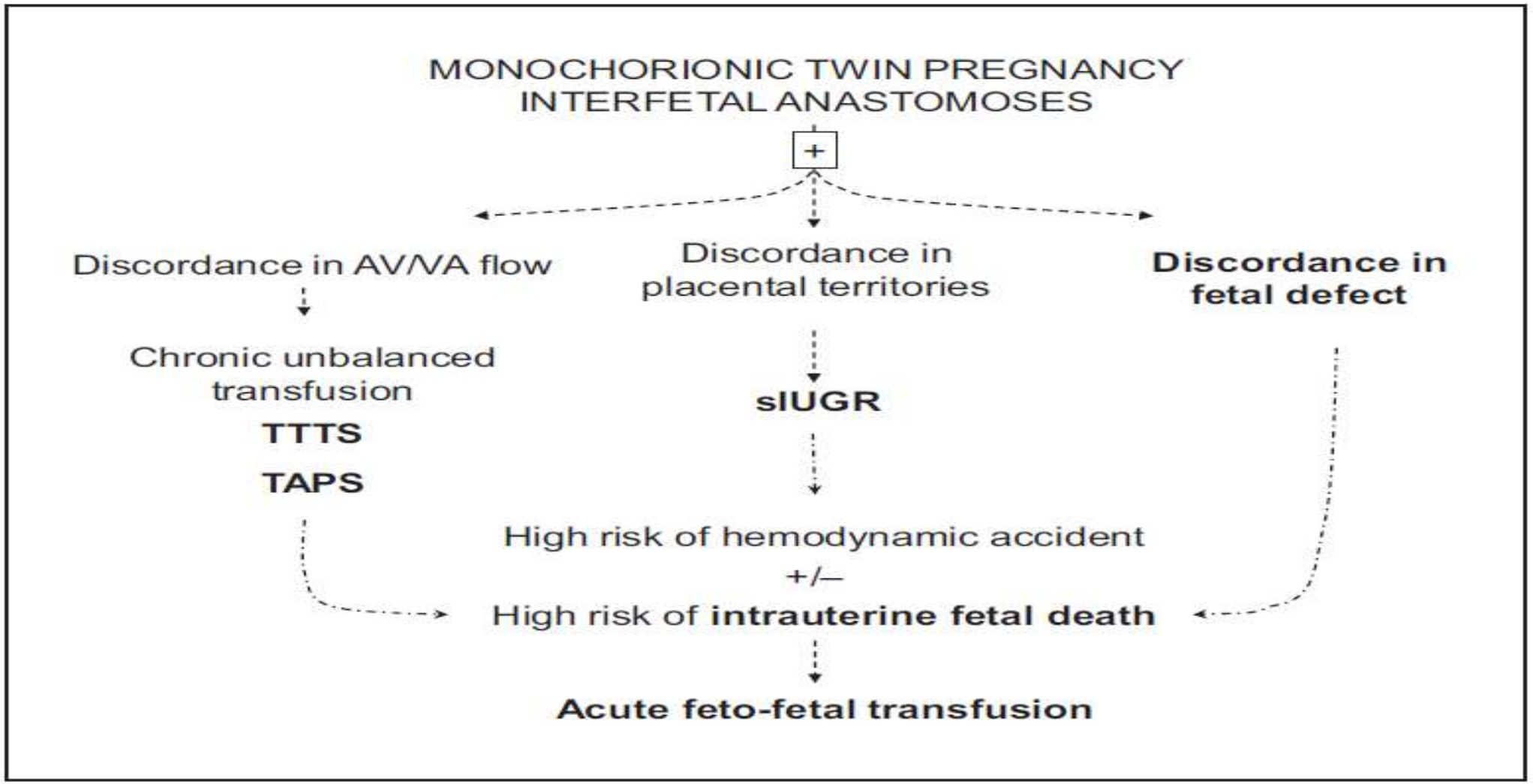
- Details: weight 450 grams (less than 3rd centile on Fenton chart)
- Pneumothorax in delivery room- ICD

- Oligohydramnios sequence
- Severe pulmonary hypoplasia

- Unfortunately could not be saved and expired on DOL-4 (pulmonary hemorrhage)

Discussion

Complications in MCDA pregnancies



Twin-to-twin transfusion syndrome (TTTS)

- Occurs in monochorionic (MCDA) multiple pregnancies
- MC twins share a single placenta and almost all cases have vascular anastomoses on the chorionic plate connecting the two fetal umbilical circulations.
- 3 different types of anastomoses: arterio-venous (in both directions-most common), arterio-arterial and veno-venous (type, number and diameter determine risk profile)

Incidence

- Approximately 10-15% of all MC twins develop TTTS which usually occurs between the 16th and 26th weeks of gestation

Diagnosis and staging of TTTS

ISUOG Practice Guidelines

- Sonographic screening for TTTS should be performed every 2 weeks from 16 weeks onwards.
- Severe amniotic fluid discordance is the main prenatal finding.
- The recipient fetus shows an increasing polyhydramnios defined as a deepest vertical pocket >8 cm before 20 weeks and >10 cm after 20 weeks of gestation (Eurofoetus group)
- The donor shows oligo or anhydramnios with a deepest vertical pocket <2 cm and is stuck within its membranes to the uterine wall or placenta by the excessive polyhydramnios of the recipient.

Quintero chart

Table 1

Staging of severe TTTS based on ultrasound findings (modified from Quintero [10]).

Stage	Poly-/Oligohydramnios	Absent bladder in donor	[a]Severely abnormal Doppler findings in UA and/or DV	Hydrops	Demise
I	+	−	−	−	−
II	+	+	−	−	−
III	+	+	+	−	−
IV	+	+	+	+	−
V	+	+	+	+	+

[a] Defined as at least one of the following: umbilical artery (UA) absent or reversed enddiastolic flow (ARED), negative A-wave ductus venosus (DV).

Approach to management of TTS

- Expectant management
- Fetoscopic laser ablation of anastomotic vessels (Solomon technique)
- Amnioreduction
- Selective fetal reduction (rarely performed in the absence of discordant malformations or severe FGR)

Fetoscopic laser surgery

Table 3

Pros and cons regarding early and late laser surgery in TTTS (modified from Baud et al. [33]).

	Early TTTS (<16 weeks)	Late TTTS (>26 weeks)
Prevalence	2.5%	4–8%
In favour of laser surgery	- Feasible - Perinatal outcome comparable to conventional laser therapy between 16 and 26 weeks	- Delay delivery and recovery in utero - Trend for better neonatal outcome
Against laser therapy	- Amnion–chorion not fused - Increased risk of PPROM rate within 1 week of laser - Hypothetic spontaneous regression	- Turbid amniotic fluid - Larger placental vessels, difficult to coagulate

Preterm management in the GOLDEN HOUR

- Antenatal steroid and magnesium sulfate administration
- Delivery at tertiary care centre
- Delayed cord clamping
- Hypothermia management
- RDS management
- Blood sugar management
- Fluid and electrolytes management
- Asepsis

Modified Ross Heart Failure Classification for children

- Class I: Asymptomatic
- Class II: Mild tachypnea or diaphoresis with feeding in infants, dyspnea on exertion in older children
- Class III: Marked tachypnea or diaphoresis with feeding in infants, marked dyspnea on exertion, prolonged feeding times with growth failure
- Class IV: Symptoms such as tachypnea, retractions, grunting or diaphoresis at rest

Goals of management of CHF in neonates

- A&E/Initial care: Ensure hemodynamic stability
  Ensure euvolemia
  Protect end organ function

- Early therapy: Prevent disease progression
  Arrest maladaptive neurohormonal pathway response
  Provide psychosocial support to the family
  Refer to higher centre with facilities for
  a. inotropic support
  b. mechanical ventilation
  Palliative care if applicable

- Long term: Ensure adequate growth and nutrition

Take Home Messages

- Identification of complication through regular antenatal ultrasounds
- Referral for fetoscopic laser surgery to a tertiary care centre if feasible at the earliest
- Antenatal corticosteroid
- Magnesium sulfate
- Stabilization within the golden hour of life
- Early surfactant administration, repeat dose if necessary in case of persistently high ventilatory requirements
- CHF management