Twin to Twin Transfusion Syndrom

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Abstract

Increased mortality in monochorionic fetus caused by vascular anastomosis in the placenta that causes Twin to Twin Transfusion Syndromes (TTTS). Reporting two cases experienced and taken care in our hospital. Case 1: a woman 28 years old was diagnosed with MP3 gravida preterm pregnancy 33-34 weeks with Twin to Twin Transfusion Syndromes. Ultrasound impression: Gemelli, gravid 33-34 weeks with TTTS. The patient was terminated by SCTPP. The first baby was female, born with: BW: 1400 grams, BL: 44 cm, A/S: 8/9, the second baby: female, BW: 1000 grams, BL: 38 cm, A/S: 6/7 the first baby Hemoglobin was 16.9 g/dl and the second baby was 11.3 g/dl. The babies were treated in Perinatology, the first baby survive with some congenital abnormalities such as VSD and Hemangioma Orbita. The second baby died at the age of 7 days with suspected sepsis. To date the baby still has a routine medical checkup to RS. DR. M. Djamil Padang. Case 2: a woman 31 years old was diagnosed with MP5 parturient aterm kala II with gemelli. The babies were born spontaneously. The first baby: BW: 3200 grams, PB: 48cm, A/S: 8/9, Bayi ke II perempuan : BB: 2100 grams, PB: 44 cm, A/S: 7/8. Kadar Hb bayi I : 17,6 g/dl dan bayi II: 14,1 g/dl. Plasenta lahir secara spontan lengkap 1 buah, berat 1150 gr, ukuran 20x19 x3 cm, panjang kedua tali pusat masing-masing 60 cm, insersi paracentralis,Monokhorion-Monoamnion. Kesan: TTTS.

Keywords: twin to twin transfusion syndrome, ultrasonografi, monokorion

Case Report

Case 1:
Reported a patient 28 years old, MP3 (Multy Parous 3) preterm pregnancy 33-34 weeks with TTTS who treated in obstetric department, DR. M.Djamil general hospital Padang on February 13, 2013. From the laboratories finding, obtained that Hemoglobin: 10.5 g/dl, leucocytes: 11.800/mm³, platelets: 231.000/mm³. From the Ultrasonography (USG) finding, Fetus I: female, BPD (Biparatal Diameter): 84mm, AC (Abdominal Circumference): 290mm, FL (Femur Length): 63mm, HL (Humerus Length): 55mm, EFW (Estimated Fetal Weight): 2161 gr, SDAU (Systolic Diastolic Artery Umbilical): 1.80, polyhydramnion with AFI (Amniotic Fluid Index) single pocket 15.5 cmbladderuk 4.6x3,0x3,4cm. Fetus II:
female, BPD: 72 mm, AC: 251 mm, FL: 60 mm, HL: 51 mm, EFW: 1317 g, 2.11S DAU severe oligohydramnion, AFI<2 cm, Stuck twin, empty bladder. Shown "T" sign.

USG on Februari 14th, 2013

Figure 1. Discordance twin: Twins showing a marked difference in size (greater than 10% in weight) at birth. The condition is usually caused by over perfusion of one twin and under perfusion of the other. It is fairly common in identical twins but may also occur in dizygotic twins.

BPD (I): 84mm, BPD (II): 72 mm

Figure 2. Polyhydramnion: excess of amniotic fluid, usually exceeding 2000 mL. AFI Single Pocket: 15.5 cm

Figure 3. Fetal bladder in donor twin is empty and fetal bladder in recipient twin is over distecy

The patient was treated at the obstetric room with Expectative management, for 7 days maintain ability maternal and fetal condition monitored closely. On February 20, 2013 an ultrasonography finding: Fetus I, Head presentation, fetal movement activity was good, Biometry: BPD: 84mm, AC: 276mm, FL: 65mm, HL: 58mm, EFW: 2161gr, SDAU: 2.66, AFI single pocket 12.2 cm, bladders uk 2.0 x 2.9 x 2.8 cm. Fetus II, attached to the front wall of the uterus (fixed/stuck twin) severe oligohydramnion. Biometry BPD: 70mm, AC: 218mm, FL: 50mm, HL: 56mm. EFW: 1317gr, SDAU: 3.00, AFI <2 cm, the bladder is empty. The placenta is embedded in the front of the corpus grade I-II. Impression: Gamely, gravid 34-35.

weeks with TTTS. Advice: SC.

USG on Februari 20th, 2013

On February 20, 2013 at 15:10 pm, SCTPP was perform, baby I: was born female, BW (Birth Weight): 1400g, BL (Birth Length): 44cm, A/S: 8/9, Baby II: was female with BW: 1000 g, BL : 38 cm , A/S (Apgar/Score): 6/7. Placental weight: 800 g, size 25x24x3 cm. Umbilical cord I length: 43 cm insertion: paracentralis, umbilical cord II length: 30cm, insertion velamentosa. There was a vascular anastomosis in the fetal placenta, impression: Monochorion-diamion, with TTTS. Fetal I Hb: 16.9 g/dl and fetal II Hb: 11.3. g/dl Both infants were treated at the Perinatology. The first baby was treated for 13 days. She had been survived with several congenital abnormalities, such as: VSD and haemangioma orbita. The second baby died at the age of 7 days due to septic.

Case 2:

Reported a patient, 31 years old was treated at the Obstetric room. Dr. M. Djamil General Hospital, Padang on January 26, 2014 with diagnosis: MP 5 second stage term parturient with Gamely. From the laboratory tests finding, obtained Hemoglobin: 10.7 g/dl, leukocytes: 5.300/mm3, platelets: 187.000/mm3. Ultrasound examination could not be performed because the patients in the second stage of labour. Baby I was born spontaneously, with BW: 3200 g, BL: 48 cm, A/S: 8/9, female. Baby II: BW: 2100 g, BL: 44 cm, A/S: 7/8, female. The placenta was born spontaneously complete 1 piece, weight:1150 g, size: 20x19x3 cm, both length of each cord 60 cm, paracentralis insertion, and impression: Monochorion Diamnion. Hb babyI:17.6 g/dl, Hb baby II: 11.4 g/dl, Ht baby I: 53%, Ht baby II: 43%, Impression: TTTS. Until now,both babies are healthy and the congenital anomalies had not been found in both infants.

Figure 4. Baby in case 2 with Discordance Twin : difference in size greater than 10% in weight at birth

Figure 5. Monochorion Diamnion Placenta : a chorion with two amniotic sac
Discussion

Approximately one-third gamely monozygotic has 2 twin monozygotic 2 amnions, 2 chorionics, and 2 placentas, sometimes 2 placentas into one piece. This situation is difficult to differentiate the dizygotic twin. The other two-thirds have one placenta, one chorionic and 1 or 2 amnions.3,4

Figure 6. Mechanism of monozygotic twinning. Black boxing and blue arrows in columns A, B, and C indicates timing of division. A. At 0 to 4 days postfertilization, an early conceptus may divide into two. Division at this early stage creates two chorions and two amnions (dichorionic, diamnionic). Placentas may be separate or fused. B. Division between 4 to 8 days leads to formation of a blastocyst with two separate embryoblasts (inner cell masses). Each embryoblast will form its own amnion within a shared chorion (monochorionic, diamnionic). C. Between 8 and 12 days, the amnion and amnionic cavity form above the germinal disc. Embryonic division lead to two embryos with a shared amnion and shared chorion (monochorionic, monoamnionic). D. Differing theories explain conjoined twin development. One describes an incomplete splitting of one embryo into two. The other describes fusion of a portion of one embryo from a monozygotic pair onto the other.3

The developmental mechanisms underlying monozygotic twinning are poorly understood. Minor trauma to the blastocyst during assisted reproductive technology (ART) may lead to the increased incidence of monozygotic twinning observed in pregnancies conceived in this manner.3,6

The outcome of the monozygotic twinning process depends on when division occurs. If zygotes divide within the first 72 hours after fertilization, two embryos, two amnions, and two chorions develop, and a diamnionic, dichorionic twin pregnancy evolves. Two distinct placentas or a single, fused placenta may develop. If division occurs between the fourth and eighth day, a diamnionic, monochorionic twin pregnancy results. By approximately 8 days after fertilization, the chorion and the amnion have already differentiated, and division results in two embryos within a common amnionic sac, that is, a monoamnionic, monochorionic twin pregnancy. Conjoined twins result if twinning is initiated later. It has long been accepted that monochorionicity incontrovertibly indicated monozygoity. Rarely, however, monochorionic twins may in fact be dizygotic (Souter and colleagues, Mechanisms for this are speculative, and zygotic manipulations that accompany ART have been implicated.3,5,6

Variations of pregnancy outcomes depend on how deep the relationship of the anastomose of the fetus blood vessel. It is related to the severity of the symptoms of TTTS.

Potential predictor for TTTS is the measurement of nuchal translucency (NT) at the early gestation, membrane fold between the ages of 15 and 17 weeks gestation, searching the arterio arterial anastomosis and umbilical cord insertion. Increased NT is a marker of chromosome anomaly, cardiac defects and genetic syndromes disorders. In monochorionic twin, an increased NT may indicate early cardiac dysfunction caused by hypervolemia congestion on recipient and is associated with subsequent development of TTTS. Clinical signs are more promising for predicting TTTS is the thickness of the membrane (membrane folding) intertwin at 15-17 weeks’ gestation, contained in monochorionic twin. This sign is believed to be a reflection of oliguria and reduced amniotic fluid in fetal donor sac, and is associated with increased of development next TTTS. Another marker for the prediction of TTTS is no arterioarterial anastomosis. Arterioarterial anastomosis can be detected by color flow mapping and pulsed Doppler since 12 weeks pregnancy.

The presence or absence of reversed end-diastolic flow in the donor umbilical artery, abnormal recipient pulsation band venous system and the absence of arterial arterial anastomosis was identified freely to predict poor outcome. Staging system based on the findings of ultrasonography and fetal outcomes (Figure 2.) include: Stage I is polyhydramnion (maximum vertical pocket ≥ 8 cm) oligohydramnion (maximum vertical pocket < 2 cm) with donor fetal bladder is still visible. Instage II, the donor fetal bladder to be empty (stuck twin). Progression to stage III occurs with severe abnormal Doppler results: absent or reversed end-diastolic flow umbilical artery Doppler fetal donor or sign of abnormal veins on recipient, such as reverse flow in the ductus venosus or pulsatile umbilical venous flow. Stage IV means fetal hydrops, and at the end of stage V corresponding to the death of one or both of the twins. The purpose of this staging system is a valuable step in the standard approach to TTTS and to prove the usefulness of counseling and individualization therapy.3,5,9

The Quintero Stagging of Twin to Twin Transfusion syndrome.5

http://jurnal.fk.unand.ac.id
Table 1. Quintero Staging System of TTTS

<table>
<thead>
<tr>
<th>Stage</th>
<th>Ultrasound Parameter</th>
<th>Categorical Criteria</th>
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<tbody>
<tr>
<td>I</td>
<td>MVP(Maximal Ventricle Pocket) of Amniotic Fluid Index</td>
<td>MVP &lt; 2 cm in donor sac, MVP &gt; 8 cm in recipient sac</td>
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<tr>
<td>II</td>
<td>Fetal Bladder</td>
<td>Non Visualization of fetal bladder in donor twin over 60 minute of observation</td>
</tr>
<tr>
<td>III</td>
<td>Umbilical Artery, ductus venosus, Umbilical vein doppler waveforms</td>
<td>Absent or reverse artery umbilical diastolic flow, reverse ductus venosus, a wave flow pulsatile umbilical vein flow</td>
</tr>
<tr>
<td>IV</td>
<td>Fetal Hydrops</td>
<td>Fetal hydrops in one or both twins</td>
</tr>
<tr>
<td>V</td>
<td>Absent fetal cardiac Activity</td>
<td>Fetal demise in one or both twin</td>
</tr>
</tbody>
</table>

Figure 2. Ultrasound finding in TTTS

Stage I, polyhydramnion (maximum vertical pocket ≥ 8cm) oligohydramnion (maximum vertical pocket < 2 cm) with donor fetal bladder is still visible; Stage II, Stuck twin with an empty bladder; Stage III, abnormal Doppler measurements (absent or reverse umbilical artery end diastolic flow, or reverse blood flow ductus venosus, or pulsatile venous blood flow); stage IV, fetal hydrops; Stage V (not shown) death of one or both twins. 4,5,8

Mortality of untreated TTTS more than 80%.

Three common treatments for TTTS at mid trimester are amnio reduction, fetoscopic laser occlusion of the vessels chorioangiopagus (floc) of vascular anastomoses and septostomy. Serial Amnio reduction can control the volume of amniotic fluid. There is variability in drainage engineering, especially the amount of fluid drainage; fast or slow drainage. Denbow dkk made aggressive Amnio drainage with 1 literat the AFI increased every 10 cm, to AFI less than 25 cm. Based on the data, amnio reduction only effective in cases of moderate TTTS, with one of the three treatment failure cases and more than half of the patients who treated are still death, one or even both fetuses. Another therapy was fetoscopic laser occlusion of the vessels chorioangiopagus (Floc) with Coagulated all locked vascular anastomosis through the fetoscopy.3,9,10

In first case, the criteria for the diagnosis of TTTS is monochorionic placenta, polyhydramnios and distended bladder in the recipient fetus and severe oligohydramnion or fetal donor anhidramnion, it seen in the presence of a very small bladder or empty and stuck twin. According to The Quintero staging, the first case includes in stage II of TTTS with Polyhydramnion in the recipient fetus and severe oligohydramnion and stuck twin in donor.

In the Second case the diagnosis of TTTS Postpartum enforced by the presence of discordance Twin, same gender, monochorionic, and differences in hemoglobin greater than 5 g/dl with anemia in smaller infants.

The differences in both cases are based on gestational age and fetal outcomes. These variations were determined by the severity of relationship between anastomose of the blood vessels. Its also related to the severity of the symptoms of TTTS.

TTTS is cause due to unidirectional deep arteriovenous (AV) anastomoses with the superficial shortcomings. The superficial anastomosis responsible for acute transfusion complications that can lead to IUFD one of fetuses. This evidence increases the likelihood of the risk of monochorionic fetus depends on the type and size of vascular flow anastomoses. Hipovolemia, oliguria and oligohydramnion occurs in the donor fetus, occurring phenomenon 'stuck twin'. Hypervolemia, polyuria and polyhydramnion occurs in the fetus recipient, and often lead to excess circulating volume and hydrops.3

Ultrasoundography diagnostic of these syndromes are the single placental mass and the absence of the lambda sign or look alike “T sign” at the end of first trimester, and oligohydramnion or anhidramnion of the donor twin (often in the absence of the bladder) and hydramnion and an enlarged recipient bladder at the second trimester. May also be discordant between fetal growth or hydrops in one or both fetuses.3

Conclusion

TTTS is caused due to unidirectional deep arteriovenous (AV) anastomoses with the superficial shortcomings. Hypovolemia, oliguria and oligohydramnion occurs in the donor fetus, occurring
phenomenon 'stuck twin'. Hypervolemia, polyuria and polyhydramnios occurs in the recipient fetus, and often lead to excess circulating volume and hydrops. If the syndrome is not treated, the survival ranges between 4% to 75%.

**Literature**