Rapid Resolution of Polyhydramnios Foretells Circulatory Collapse for the Donor Twin in Feto-Fetal Transfusion Syndrome

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Feto-fetal transfusion syndrome is a pathological process unique to diamniotic monochorionic pregnancies. It is the consequence of an unbalanced fetal blood flow through communicating vessels within a shared placenta. When it occurs, a polyuric, hypervolemic recipient twin co-exists with a hypovolemic oliguric donor. The presence of polyhydramnios or oligohydramnios is considered a poor prognostic indicator, whereas normal amniotic fluid volumes indicate a lack of clinically significant twin-twin transfusion. In addition, the spontaneous normalization of amniotic fluid volume is usually seen as a favorable prognostic sign. Here, however, we present a case of feto-fetal transfusion in a 31 year-old primigravida at 19 week, in which the spontaneous normalization of amniotic fluid volume in the recipient twin preceded the death of the donor. [*P R Health Sci J 2016;35:43-45*]

Key words: Monochorionic twins, Feto-fetal transfusion syndrome, Polyhydramnios, Oligohydramnios

The implications of monochorionic twin pregnancies are significant because fetuses and neonates of monochorionic gestations account disproportionately to the overall rates of adverse neonatal outcomes and health care expenditures. In particular, monochorionicity confers a higher risk for perinatal morbidity and mortality, mainly as a consequence of vascular connections within the placenta that results in feto-fetal transfusion.

Feto-fetal transfusion syndrome is a pathological process unique to diamniotic monochorionic pregnancies. It is the consequence of an unbalanced fetal blood flow through communicating vessels within a shared placenta (1-4). When it occurs, discordant amniotic fluid volumes dominate the clinical picture. In addition, a hazardous intrauterine environment develops in which a polyuric, hypervolemic recipient twin co-exists with a hypovolemic oliguric donor. Over time, recipient twins can develop severe cardiac dysfunction leading to fetal hydrops. Because frequent antenatal testing may improve perinatal outcomes, accurate determination of chorionicity is fundamental and should be determined promptly and accurately (5-6).

Based on sonographic and clinical parameters, Quintero and his group developed a staging classification system of feto-fetal transfusion syndrome (7) (Table 1). According to their scheme, the presence of polyhydramnios or oligohydramnios is considered a poor prognostic indicator, whereas normal amniotic fluid volumes indicate a lack of clinically significant twin-twin transfusion.

The only intervention that actively targets the pathophysiology of feto-fetal transfusion syndrome is laser ablation, whose goal is to functionally separate the placenta into two independent organs. Even with this procedure, intact survival of both twins is approximately 50% (8-9). The control of amniotic fluid volumes by serial amniocenteses has success rates similar to those of laser photocoagulation and should be considered when laser ablation is not available (10).

The spontaneous normalization of amniotic fluid volume is usually seen as a favorable prognostic sign. Here, however, we present a case of feto-fetal transfusion in which the spontaneous normalization of amniotic fluid volume in the recipient twin preceded the death of the donor.

Case Report

A 30-year-old primigravida with a spontaneous twin pregnancy was referred for consultation from an outside institution. Initial ultrasound examination revealed a 19-week monochorionic diamniotic twins gestation. Fetus A had mild ventriculomegaly, growth restriction and oligohydramnios (maximum vertical pocket of less than 2 cm). Umbilical artery Doppler studies demonstrated absent end diastolic flow. Fetus B had polyhydramnios (maximum vertical pocket of 14 cm) and

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a normal umbilical artery Doppler study. These findings were consistent with the diagnosis of feto-fetal transfusion syndrome. Fetal echocardiography was performed and confirmed normal cardiac structure and hemodynamic function on both twins.

The family was counseled regarding the ultrasound findings, the natural course of the disease and the available treatment options. The patient decided against intervention.

Ultrasound follow-up at 27 weeks demonstrated adequate growth in the recipient twin with a resolution of the polyhydramnios. There was worsening growth restriction in the donor with no improvement in the oligohydramnios.

At 29 weeks gestational age, intrauterine death of the donor twin was diagnosed and the patient was admitted to the hospital for continued surveillance.

At 34 weeks gestation, labor was induced. A female neonate weighing 2100 grams was delivered. Apgar scores were 8 and 9 at 1 and 5 minutes, respectively. The neonate was discharged from the hospital after two weeks in good health and without significant complications. Pediatric follow-up at 8 months showed no clinical or radiological evidence of neurologic damage in the child.

Written informed consent was obtained from the patient for publication of this case report.

Table 1. Staging classification system of feto-fetal transfusion syndrome (7)

Stage	Sonographic features	Description
I	Amniotic fluid volume	Discrepancy between the two amniotic sacs. Polyhydramnios (maximal vertical pocket >8 cm) in recipient sac and oligohydramnios (maximal vertical pocket <2 cm) in donor sac.
II	Fetal bladder	Donor twin bladder no longer visible. Subjected to continuous observation over at least an hour.
III	Fetal Doppler waveforms	Critical abnormal values (Absent or reversed flow umbilical artery diastolic flow, reversed ductus venosus a-wave flow, pulsatile umbilical vein flow).
IV	Fetal hydrops	Abnomal collection of fluid in at least two different fetal organ spaces (in one or both twins).
V	Fetal demise	Absent cardiac activity of either fetus.

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Discussion

In 1882, the German obstetrician Friedrich Schatz observed that all monochorionic placentas contained vascular anastomoses linking the circulations of the fetuses. More importantly, he noted that if these anastomoses were not balanced in number or were discordant in size, feto-fetal transfusion syndrome occurred (11). One hundred years later, in a landmark study, Bajorja and colleagues confirmed these observations. Through dye-contrast injection, they delineated the circulation of 20 monochorial placentas from pregnancies with and without evidence of feto-fetal transfusion. They concluded that placental vascular anastomoses in monochorial pregnancies complicated by feto-fetal transfusion syndrome are both, fewer in number and of a different type than those without the syndrome (80% vs. 36% in controls, $p < 0.01$) (12). Recent literature suggests that beyond placental angioarchitecture and transfusion unbalance, other factors such as renin-angiotensin system activation in the fetoplacental unit are involved in the pathophysiology of this condition (13).

According to Quintero and his group, increased amniotic fluid volume in the recipient accompanied by oligohydramnios in the donor, is associated with an increased risk for adverse outcomes. Conversely, spontaneous normalization of amniotic fluid volume is generally perceived as an encouraging sign because it suggests that chronic hypervolemia has abated (7).

A prospective study to validate the Quintero staging system was performed by Taylor et al. using 52 cases of feto-fetal transfusion syndrome. The survival rates results were 58%, 60%, 42%, 43%, and 0% for stages I–V, respectively (14). They concluded that the Quintero staging system did not predict survival either at the time of presentation or at first treatment. Although convenient to describe ultrasound findings, its poor ability to predict disease progression or to stratify risk for adverse outcomes illustrates the limitations of this system.

In our case report, the rapid and spontaneous normalization of amniotic fluid volume in the recipient twin without improvement in the oligohydramnios of the donor, may have represented an ominous sign of cardiovascular failure rather than a "return to normal". This would decrease the amount of blood transferred to the recipient fetus causing a reduction in amniotic fluid production. It would also portend the death of the donor twin. This case exemplifies that although convenient for describing the severity of the disease, the Quintero staging system is fraught by limitations in determining pregnancy outcomes, and thus, should be used cautiously.

Resumen

El síndrome de transfusión feto-fetal es un proceso patológico reflejado únicamente en embarazos diamnióticos monocoriónicos. Es la consecuencia de un flujo de sangre fetal no balanceado a través de vasos sanguíneos dentro de una placenta compartida. Cuando ocurre, un receptor gemelo poliúrico hipervolémico coexiste con un donante hipovolémico oligúrico. La presencia de polihidramnios u oligohidramnios es considerado un indicador diagnóstico pobre, mientras que volúmenes de fluido amniótico normal indica una falta clínicamente significativa de transfusión de gemelo a gemelo. Además, la normalización espontánea de volumen de fluido

amniótico generalmente es vista como una señal de diagnóstico favorable. Aquí, sin embargo, presentamos un caso de transfusión feto-fetal en una primigrávida de 31 años de edad en la semana 19, en la que la normalización espontánea del volumen de fluido amniótico en el gemelo receptor precedió la muerte del donante.

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