Case Report of Autopsy and Placental Examination After Radiofrequency Ablation of an Acardiac Twin

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ABSTRACT
We report the autopsy and placental findings in a monochorionic twin gestation complicated by twin reversed arterial perfusion (TRAP) sequence. Radiofrequency ablation (RFA) was performed at 24 weeks gestation to abort the acardiac fetus, and vaginal delivery of the co-twin and acardiac fetus occurred at 33 weeks gestation. An autopsy of the acardiac fetus revealed multiple congenital anomalies including complete absence of the upper extremities and poor development of the skull and facial structures. In contrast to the upper body, the lower half of the body, although malformed, was more developed. The monochorionic twin placenta showed velamentous, atrophied, proximal artery-artery and vein-vein intertwin vascular connections which essentially bypassed the placental parenchyma for the acardiac fetus. Ink injection and histologic examination confirmed thrombosis of these critical intertwin vascular connections after RFA. This report highlights the fetal and placental anatomy of TRAP sequence and stresses the importance of placental examination after fetal surgical techniques.

Keywords: fetal laser coagulation, pathophysiology, monochorionic twin, acardiac twin, laser ablation, twin reversed arterial perfusion (TRAP)

Abbreviations:
TTTS, twin-to-twin transfusion syndrome; TRAP, twin reversed arterial perfusion; A-A, artery-artery; V-V, vein-vein; RFA, radiofrequency ablation; A-V, arteriovenous

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Case Report

A 24-week-old acardiac African American fetus (twin B) of a 21-year-old African American woman designated...
gravida 2 para 1 (in her second pregnancy, with previous pregnancy having been spontaneously or electively aborted) delivered a healthy son (twin A) at 33 weeks and 5 days gestation. The pregnancy had been a monochorionic twin gestation with TRAP sequence which was discovered via an ultrasound examination performed at 22 weeks, when the mother arrived at our facility for prenatal care. Subsequently, the acardiac fetus (twin B) was selectively aborted using radiofrequency ablation (RFA) at 24 weeks gestation. Amniocentesis of twin A was performed, which revealed a normal karyotype. Twin A was followed up with weekly ultrasound examinations and subsequently was born via normal, spontaneous vaginal delivery at 33 weeks and 5 days, after preterm premature rupture of the extraplacental membranes.

After delivery, an autopsy of twin B was requested, which revealed a moderately macerated fetus (sex indeterminate), small for gestational age, with a body weight of 125 g (expected weight at 24 weeks gestation, mean [SD] 586[74] grams). The fetus exhibited multiple congenital anomalies, which are demonstrated and summarized in Image 1. Most notably, there was complete absence of the upper extremities and poor development of the skull and facial structures. In contrast to the upper body, the lower half of the body, although malformed, was more developed, consistent with the TRAP sequence and acardiac-twin phenomena.

We received the twin placenta which was submitted for pathologic examination. The focally disrupted monochorionic, diamnionic twin placenta weighing 370 g, had a thin, transparent dividing membrane and only 1 definitive umbilical cord. The extraplacental membranes showed an atrophied velamentous umbilical artery (0.1 cm in diameter) and a velamentous umbilical vein (0.3 cm in diameter), representing likely A-A and V-V intertwin vascular connections (Image 2A). Ink injection of the umbilical vessels of twin A demonstrated a direct arterial connection between the umbilical artery of twin A and the atrophied smaller velamentous vessel. The velamentous
vein connected directly to the umbilical vein of twin A. Ink injection also confirmed significant involution of these critical intertwin vascular connections after RFA: ink failed to pass through the length of the 2 velamentous vessels associated with the acardiac fetus (Image 2B). Histologic examination of velamentous blood vessels at the placental margin connecting the acardiac twin to the placenta showed thrombosed blood vessels (Image 2B, inset). These findings confirmed that before RFA twin A perfused the acardiac twin, bypassing the placenta, and that RFA was successful.

Twin A was a preterm, vigorous male infant born at 33 weeks and 5 days, weighing 2000 g, with Apgar scores of 9 at 1 minute after birth and 9 at 5 minutes after birth. He was admitted to the neonatal intensive care unit based on his prematurity and was given gavage enteral feeds. His stay was complicated only by brief periods of bradycardia (attributed to apnea of prematurity), nasal cannula support given for low oxygen saturation, and mild jaundice that improved with phototherapy. After a 20-day hospital stay, the infant was discharged to home, with a body weight of 2235 g and normal neurological and musculoskeletal assessment with no apnea, bradycardia, or desaturations. After his discharge from the hospital, a community health care practice took over his care. Further follow-up information is not available.

**Discussion**

This report highlights the importance of careful examination of all monochorionic twin placentas and demonstrates the unusual vascular connections in TRAP sequences. Intertwin vascular connections occur in virtually all monochorionic twin placentas; 3 main types are recognized, namely, A-A, V-V, and arteriovenous (A-V). The A-A and V-V connections are direct, superficial communications between chorionic plate vessels; in vivo connections have bidirectional flow. Therefore, A-A and V-V connections can be demonstrated in the delivered placenta by the ability to move blood (or other injected liquid, dye, and/or ink) freely from the vessel associated with 1 twin to the vessel of the other on the chorionic plate. A-V connections are not superficial connections; they are considered to be deep. They are recognized on the chorionic plate of the placenta by the presence of an unpaired artery from 1 twin diving into a cotyledon with an unpaired vein from the co-twin near the vascular equator. The deep anastomosis for A-V connections occurs at the level of the villous capillary. A-V connections are, by definition, unidirectional.

Although nearly every monochorionic twin placenta has intertwin vascular connections, not all will develop TTTS;
the exact mechanisms that lead to TTTS are not fully understood. However, placental characteristics such as intertwin vascular connections, peripheral cord insertion, placental share, and chorionic vascular distribution pattern are thought to be important factors. The presence of A-V connections is probably critical for the development of TTTS; documentation of such connections is important; however, it is still not clear that an imbalance of A-V connections is required to develop TTTS. Further, the frequency of A-A connections is lower in TTTS placentas, and the frequency of V-V connections is higher in TTTS placentas. Despite this, the exact functional consequence of these superficial vascular connections and how they may promote development of or protect against TTTS is currently unknown.

Also, peripheral cord insertion (marginal or velamentous) and unequal placental sharing have been linked to risk of development of TTTS. The donor twin in TTTS is more likely to have peripheral cord insertion and/or the smaller share of the placenta. Lastly, the pattern of distribution of the chorionic plate vessels has been associated with TTTS. Specifically, the magistral pattern of vessel branching, in which a large vessel traverses from the umbilical insertion site to the periphery of the placenta without significant reduction in size or branching, is observed more commonly in placentas complicated by TTTS and is more common in the donor twin placenta. This combination of placental characteristics associated with TTTS suggests a complex and multifactorial pathogenesis for TTTS.

In healthy twins in utero, oxygen- and nutrient-enriched blood enters the placenta through the umbilical arteries (represented by the blue circle and arrow) of each twin. Blood is enriched in oxygen and nutrients and then returns to each twin through their separate umbilical veins (represented by the red arrow and red circle). The circulations, in theory, are 2 complete and separate circuits. Twin reversed arterial perfusion (TRAP) sequence. The pump twin circulation to the placenta is essentially unchanged from normal circulation. However, due to the large artery-artery connection between the pump twin and the acardiac twin, the pump twin perfuses the acardiac twin with oxygen- and nutrient-poor blood. The acardiac twin receives little or no oxygen- and nutrient-rich blood from the placenta because of the large vein-vein connection. The direction of flow in the umbilical vein and artery of the acardiac twin is reversed because of the vascular connections with the pump twin. Conceptually, this circulation is in series, rather than in parallel. Because the acardiac twin receives oxygen- and nutrient-poor blood from the pump twin and because the blood enters the fetus at the umbilicus, the lower extremities and abdomen consume the oxygen and nutrients that remain, leaving inadequate supply for the development of the upper extremities, torso, and head.
Gross examination of the fetal vessels leading to the acardiac twin showed no residual umbilical cord attached to the acardiac twin. The abdominal wall at the site of the umbilical cord insertion was disrupted, and this could have been secondary to local necrosis after RFA. We observed no residual umbilical cord attached to the acardiac twin. Gross examination of the fetal vessels leading to the cord and body of the acardiac twin appeared atrophic and thrombosed, likely secondary to lack of blood flow subsequent to the RFA.

Conclusion

In this case, intrauterine RFA was used to stop the blood flow to the acardiac twin and to ablate flow through the placental vascular connections; aborting the acardiac twin prevented the death of the pump twin. Ink injection studies of the monochorionic twin placenta, performed after delivery, confirmed involution of critical placental vascular connections after RFA.

This case report highlights the importance of placental examination after fetal surgical techniques, documents vascular connections and can contribute significantly to understanding of the pathophysiology and other complications of monochorionic twinning.

References


