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Diagnosis of twin-to-twin transfusion syndrome, selective fetal growth restriction, twin anaemia-polycythaemia sequence, and twin reversed arterial perfusion sequence

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Monochorionic twin pregnancies are well known to be at risk for a variety of severe complications, a true challenge for the maternal–fetal medicine specialist. With current standards of care, monochorionicity should be established in the first trimester. Subsequently, frequent monitoring using the appropriate diagnostic tools, and in-depth knowledge about the pathophysiology of all possible clinical presentations of monochorionic twin abnormalities, should lead to timely recognition, and appropriate management. Virtually all unique diseases found in monochorionic twins are directly related to placental angio-architecture. This, however, cannot be established reliably before birth. The clinician needs to be aware of the definitions and symptoms of twin-to twin transfusion syndrome, selective fetal growth restriction, twin anaemia–polycythaemia sequence, and twin reversed arterial perfusion sequence, to be able to recognise each disease and take the required action.

In this chapter, we address current standards on correct and timely diagnoses of severe complications of monochorionic twin pregnancies.

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Introduction

Monochorionic twin pregnancies are associated with a variety of serious complications, unique to this group. The underlying pathophysiology of most of these complications seems to be related, directly or indirectly, to the unique angio-architecture of the monochorionic placenta [1]. Imbalanced blood flow, with net transfer of blood and likely of numerous regulating factors, is the accepted cause of twin-twin transfusion syndrome (TTTS) and twin anaemia–polycythemia sequence (TAPS). Although the accepted cause of selective fetal growth restriction (sFGR) is an unequally shared placenta, and the reversed blood flow in the cord and the body of the acardiac twin explains its features, the underlying pathophysiology may start with imbalanced blood flow through particular types of vascular anastomoses. With current imaging technology, we cannot reliably visualise these anastomoses. Ultrasound and Doppler examinations enable us to detect early signs and symptoms of the various pathologic conditions in monochorionic twins. As for some, treatment options exist, timely diagnosis and referral is of vital importance. In this chapter, we aim to discuss the clinically relevant diagnostic tools that can and should apply in the management of monochorionic twin pregnancies.

Twin-to-twin transfusion syndrome

Monochorionic twins share a single placenta with vascular anastomoses that connect the fetal circulations, allowing inter-twin blood transfusion. This is a physiological phenomenon as long as blood flow between the fetuses is balanced. Unbalanced net inter-twin blood transfusion, however, may lead to various forms of serious pathology. The best-known clinical syndrome is TTTS, which is a chronic form of feto–fetal transfusion and affects about 9% of monochorionic twins [2].

In the past, TTTS was diagnosed at the time of birth based on neonatal criteria that included a growth discordance of 15–20% associated with discordant cord or neonatal haemoglobin concentration of $\geq$5 g/dl [3]. A subsequent study by Wenstrom et al. [4] showed that any combination of weight and haemoglobin discordance could be seen with equal frequency. The investigators concluded that the historical paediatric parameters should be abandoned for the diagnosis of TTTS.

Nowadays, TTTS is diagnosed prenatally by ultrasound examination. It typically presents in the second trimester of pregnancy, and the diagnosis is based on the presence of a twin oligo–polyhydramnios sequence (TOPS). The following ultrasound criteria are required to diagnose TTTS: (1) the twin gestation is diagnosed as monochorionic (ideally with a scan carried out in the first trimester showing a T-sign); and (2) a combination of oligohydramnios exists in one twin's amniotic cavity, with polyhydramnios in the other twin's amniotic cavity. Oligohydramnios is defined as a maximum vertical pocket (MVP) of $\leq$2 cm. This twin is commonly called the donor twin. Polyhydramnios was originally defined as a MVP of $\geq$8 cm [5]. Subsequent studies in Europe have proposed to increase the threshold to 10 cm or more for the MVP after 20 weeks' gestation [6]. This twin is described as the recipient twin.

A twin gestation that meets the above criteria is classified as TTTS. Quintero et al. [5], in 1999, described a classification of TTTS (Table 1), which has been widely adopted and used throughout the world.

Table 1
Staging of twin-to-twin transfusion syndrome.

Stage	Oligo/ polyhydramnios[a]	Absent bladder filling in donor	Critically abnormal Doppler studies[b]	Hydrops of either fetus	Intrauterine fetal demise of either fetus
I	+	–	–	–	–
II	+	+	–	–	–
III	+	+	+	–	–
IV	+	+	+	+	–
V	+	+	+	+	+

[a] Oligohydramnios: maximum vertical pocket of 2 cm or less; polyhydramnios: maximum vertical pocket of 8 cm or over (or $\geq$10 cm at >20 weeks' gestation).
[b] Absent or reversed diastolic velocity in the umbilical artery or ductus venosus or pulsatile umbilical venous velocity.