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Right ventricular outflow tract anomalies in monochorionic twin pregnancies diagnosed prenatally

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Objective

Right ventricular outflow tract anomalies (RVOTA) include pulmonary stenosis, atresia and insufficiency and are reported in 8-12% of the recipient twins of monochorionic twin (MC) pregnancies complicated by twin to twin transfusion syndrome (TTTS). It has been speculated that in MC pregnancies complicated by TTTS, RVOTA could be the consequence of an unbalanced passage of blood and vasoactive mediators, through the vascular anastomosis, between the donor and the recipient fetuses. However, prenatal diagnosis of RVOTA has been reported not only in recipients of untreated TTTS but also in donors, larger twin of a MC pair with selective intrauterine growth restriction (sIUGR), and as an isolated discordant anomaly. The aim of this study was to evaluate the pregnancy characteristics at diagnosis of RVOTA, and the outcomes of RVOTA observed in a large cohort of MC pregnancies managed at a single referral center.

Methods

All monochorionic pregnancies referred to our center between 2009 and 2018 were retrospectively reviewed for a prenatal diagnosis of RVOTA. RVOTA were classified according to the characteristic of the flow across the pulmonary valve (PV) and in the pulmonary artery (PA) in: pulmonary stenosis (PS), if forward flow was present across PV and in PA but with a peak systolic velocity (PSV) > 100 cm/sec; pulmonary insufficiency (PI), if bidirectional flow was present across the PV and in PA; pulmonary steno-insufficiency (PSI), if both the previous findings were present; pulmonary atresia (PAt), if no flow was present across PV and an exclusive reverse flow was present in PA. Pregnancy characteristics at the time of RVOTA diagnosis were recorded, including TTTS before fetoscopic laser surgery (FLS), TTTS treated with FLS with or without recurrence, twin anemia polycythemia sequence (TAPS) spontaneous or after FLS, sIUGR, amniotic fluid discordance (>3cm in deepest vertical pocket), isolated RVOTA as discordant anomaly. Fetuses with other structural abnormalities in addition to RVOTA were excluded. Echocardiographic assessments from prenatal RVOTA diagnosis until the neonatal period were evaluated and data on outcome were collected.

Results

During the study period, 1642 MC pregnancies were referred to our center and 38 cases of RVOTA were identified (2.3%). Pregnancy characteristics at the diagnosis of RVOTA, prenatal diagnosis and postnatal outcomes of cases observed in our cohort are summarized in Table 1.

Conclusion

Right ventricle outflow tract anomalies occur both in complicated and uncomplicated MC twin pregnancies. In those with TTTS not only the recipients, but also the donors may be affected, and the anomalies in the recipient can persist despite successful treatment. In Right ventricular outflow tract detected in utero requires postnatal cardiological follow up.