FETAL ECHO IN MULTIPLE PREGNANCIES

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Introduction

Twin gestations account for 1-2% of all pregnancies. Dyzygotic twins originate from 2 eggs and are associated with dichorionic, diamniotic pregnancies whereas monozygotic twins originate from mitotic division of one egg and may have monochorionic or dichorionic placentation.[1] Population probabilities are that 30% of twin pregnancies are monozygotic. The type of chorionicity can be determined accurately by ultrasound[2] while prediction of zygosity is less certain.[3] The outcome of multiple pregnancies is greatly affected by chorionicity whereas the effect of zygosity is less clear. Having a shared placenta increases the risk of adverse perinatal outcome thus, efforts should be made to determine chorionicity.[1]

Multiple pregnancies and congenital heart disease

In a population based study, twins of the same sex appeared to be at greater risk of congenital anomalies[4] but it was less clear if this was related to zygosity or chorionicity. More recently, Dube et al showed higher-but no statistical difference in the incidence of major anomalies when comparing monozygotic twins (mono or dichorionic) with dyzygotic pairs.[1] Congenital heart defects occur more commonly in monozygotic twins who are monochorionic than in monozygotic dichorionic twins.[5] In another study a higher incidence of congenital heart disease was shown in twins who are monochorionic diamniotic (~3.8%) compared to that of the general population. In addition, the risk appeared to be higher in those with (6.9%) than in those without (2.3%) associated twin-to-twin transfusion syndrome (TTTS).[6] Thus, there is a place to perform detailed fetal echocardiography in monochorionic pregnancies.

Following the diagnosis of a major structural abnormality in a singleton, subsequent pregnancy management may be relatively straightforward. If, however, the pregnancy involves one affected and one unaffected twin, management issues such as termination are more complex.[7] Counselling is best performed by a combined approach between the fetal cardiologist and most importantly, the obstetric team. Copel et al reviewed their experience of fetal echo in multiple gestations in order to evaluate how the presence of a cardiac defect in one twin affected decision management for the pregnancy.[8] There were 11 defects in 10 set of twins out of 36 pregnancies studied, 7 of whom were referred because of suspected heart defect. Three fetuses were aneuploid including one set of monozygotic twins with trisomy 18. Three of 8 pregnancies were monochorionic and there were no cases of selective fetocide. The information provided by fetal echocardiography allowed comprehensive family counselling leading to informed decisions regarding intrapartum and neonatal management in a similar fashion to management of a singleton pregnancy given that no termination was performed.

Of growing interest in twins pregnancies however, is the assessment of twin-to-twin transfusion syndrome which is associated with important morbidity and mortality[9;10] In addition to characterising structural cardiac abnormalities, there also appears to be an important role for fetal echocardiography in assessing cardiac function in these high-risk pregnancies.

Fetal echocardiography and twin-to-twin transfusion syndrome

TTTS is a common, severe complication of monochorionic pregnancies with a quoted incidence that varies from 5-35%[9;11;12] Its pathophysiology is not fully understood but thought to occur due to vascular connections in the placenta leading to an umbalanced circulation between the twins with shift of blood from one (the donor) to the other (the recipient)[13;14]

Most dramatic changes occur in the recipient twin and characteristically affect the right ventricle more than the left. Progression of TTTS leads to cardiomegaly, impaired cardiac function and ultimately hydrops. Variable degree of right and left ventricular hypertrophy and dilatation, tricuspid regurgitation and some left ventricular dysfunction have been reported[1] Ventricular dilatation and hypertrophy is thought to be due to increased preload. Some fetuses may show disproportionate degree of hypertrophy. Possible explanations for this may involve the effect of hormones originating in the donor twin that cross the placenta further to affect the recipient twin by increasing its afterload. Examples are the renin-angiotensin system[16;17] and endothelin-1[18] Angiotensin may also be linked to hypertension seen in the recipient twin[19] which may explain ventricular hypertrophy. Right ventricular outflow tract obstruction is also well documented in the recipient. It can progress during fetal life leading to pulmonary atresia in the most severe cases and may progress further post-natally. The obstruction may be seen at the level of the pulmonary valve and / or muscular subvalvar area.[20-22]

Characteristically and contrary to the recipient, the donor twin shows no overt cardiac disease[15;21] More recently however. left ventricular shortening fraction has been shown to be higher in the donor than in the recipient twin, possibly reflecting a response to anemia[23;24] No intertwin differences in haemodynamic parameters are expected to occur in normal twin pregnancies.[25] Raboisson and colleagues have recently shown abnormalities of diastolic function early in the course of TTTS. Myocardial performance indices (MPI, 'Tei index') were shown to be systematically higher in the recipient when compared to the donor twin. The higher MPI values encountered were due to prolongation of the isovolumic relaxation time, in keeping with diastolic abnormalities. At the same time, in the earlier phases of TTTS, indices of systolic function remained normal[24] Progressive biventricular hypertrophy (cardiomyopathy) with predominant right ventricular systolic and biventricular diastolic dysfunction have also been shown in the course of TTTS.[26] These recent studies may further substantiate the theory of increased afterload being important in the pathophysiology of TTTS.

References

1. Dube J, Dodds L, Armson BA. Does chorionicity or zygosity predict adverse perinatal outcomes in twins? Am J Obstet Gynecol 2002; 186(3):579-583.
2. Finberg HJ. The "twin peak" sign: reliable evidence of dichorionic twinning. J Ultrasound Med 1992; 11(11):571-577.
3. Scardo JA, Ellings JM, Newman RB. Prospective determination of chorionicity, amnionicity, and zygosity in twin gestations. Am J Obstet Gynecol 1995; 173(5):1376-1380.
4. Layde PM, Erickson JD, Falek A, McCarthy BJ. Congenital malformation in twins. Am J Hum Genet 1980; 32(1):69-78.
5. Cameron AH, Edwards JH, Derom R, Thiery M, Boelaert R. The value of twin surveys in the study of malformations. Eur J Obstet Gynecol Reprod Biol 1983; 14(5):347-356.
6. Karatza AA, Wolfenden JL, Taylor MJ, Wee L, Fisk NM, Gardiner HM. Influence of twin-twin transfusion syndrome on fetal cardiovascular structure and function: prospective case-control study of 136 monochorionic twin pregnancies. Heart 2002; 88(3):271-277.
7. Blickstein I. Controversial issues in the management of multiple pregnancies. Twin Res 2001; 4(3):165-167.
8. Copel JA, Inati M, Green JJ, Hobbins JC, Keinman CS. Fetal echocardiography in multiple gestations. Ultrasound Obstet Gynecol 1991; 1(2):111-114.
9. Urig MA, Clewell WH, Elliott JP. Twin-twin transfusion syndrome. Am J Obstet Gynecol 1990; 163(5 Pt 1):1522-1526.
10. Burke MS. Single fetal demise in twin gestation. Clin Obstet Gynecol 1990; 33(1):69-78.
11. Blickstein I. The twin-twin transfusion syndrome. Obstet Gynecol 1990; 76(4):714-722.
12. Duncan KR, Denbow ML, Fisk NM. The aetiology and management of twin-twin transfusion syndrome. Prenat Diagn 1997; 17(13):1227-1236.
13. Bajoria R, Wigglesworth J, Fisk NM. Angioarchitecture of monochorionic placentas in relation to the twin-twin transfusion syndrome. Am J Obstet Gynecol 1995; 172(3):856-863.
14. De Lia J, Fisk N, Hecher K, Machin G, Nicolaides K, Hyett J et al. Twin-to-twin transfusion syndrome--debates on the etiology, natural history and management. Ultrasound Obstet Gynecol 2000; 16(3):210-213.
15. Fesslova V, Villa L, Nava S, Mosca F, Nicolini U. Fetal and neonatal echocardiographic findings in twin-twin transfusion syndrome. Am J Obstet Gynecol 1998; 179(4):1056-1062.

16. Mahieu-Caputo D, Dommergues M, Delezoide AL, Lacoste M, Cai Y, Narcy F et al. Twin-to-twin transfusion syndrome. Role of the fetal renin-angiotensin system. Am J Pathol 2000; 156(2):629-636.
17. Mahieu-Caputo D, Muller F, Joly D, Gubler MC, Lebidois J, Fermont L et al. Pathogenesis of twin-twin transfusion syndrome: the renin-angiotensin system hypothesis. Fetal Diagn Ther 2001; 16(4):241-244.
18. Bajoria R, Sullivan M, Fisk NM. Endothelin concentrations in monochorionic twins with severe twin-twin transfusion syndrome. Hum Reprod 1999; 14(6):1614-1618.
19. Mahieu-Caputo D, Salomon LJ, Le Bidois J, Fermont L, Brunhes A, Jouvet P et al. Fetal hypertension: an insight into the pathogenesis of the twin-twin transfusion syndrome. Prenat Diagn 2003; 23(8):640-645.
20. Zosmer N, Bajoria R, Weiner E, Rigby M, Vaughan J, Fisk NM. Clinical and echographic features of in utero cardiac dysfunction in the recipient twin in twin-twin transfusion syndrome. Br Heart J 1994; 72(1):74-79.
21. Lougheed J, Sinclair BG, Fung Kee FK, Bigras JL, Ryan G, Smallhorn JF et al. Acquired right ventricular outflow tract obstruction in the recipient twin in twin-twin transfusion syndrome. J Am Coll Cardiol 2001; 38(5):1533-1538.
22. Simpson LL, Marx GR, Elkadry EA, D'Alton ME. Cardiac dysfunction in twin-twin transfusion syndrome: a prospective, longitudinal study. Obstet Gynecol 1998; 92(4 Pt 1):557-562.
23. Lachapelle MF, Leduc L, Cote JM, Grignon A, Fouron JC. Potential value of fetal echocardiography in the differential diagnosis of twin pregnancy with presence of polyhydramnios-oligohydramnios syndrome. Am J Obstet Gynecol 1997; 177(2):388-394.
24. Raboisson MJ, Fouron JC, Lamoureux J, Leduc L, Grignon A, Proulx F et al. Early intertwin differences in myocardial performance during the twin-to-twin transfusion syndrome. Circulation 2004; 110(19):3043-3048.
25. Sonesson SE, Fouron JC, Leduc L, Lessard M, Grignon A. Reference values for differences between cardio-circulatory variables of normal twin fetuses. Ultrasound Obstet Gynecol 2000; 15(5):407-412.
26. Barrea C, Alkazaleh F, Ryan G, McCrindle BW, Roberts A, Bigras JL et al. Prenatal cardiovascular manifestations in the twin-to-twin transfusion syndrome recipients and the impact of therapeutic amnioreduction. Am.J.Obstet.Gynecol. 192, 892-902. 2005.