CONGENITAL HEART DISEASE

Atrioventricular and ventriculoarterial discordance (congenitally corrected transposition of the great arteries): echocardiographic features, associations, and outcome in 34 fetuses

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Objective: To identify fetal echocardiographic features, associations, and outcome of atrioventricular and ventriculoarterial discordance.
Design: Retrospective analysis of videotape recordings of fetal echocardiograms and examination of departmental database and patient records to establish outcome.
Setting: Tertiary centre for fetal cardiology.
Patients: 34 consecutive fetuses with a prenatal diagnosis of double discordance seen between January 1993 and December 2003.
Main outcome measures: Echocardiographic features including reversed differential insertion, identification of the right ventricle through the moderator band, and the origin and orientation of the great arteries. All associated cardiac abnormalities and outcome.
Results: Reversed differential insertion was identified in 26 fetuses. The right ventricle could be identified by the moderator band in 30 fetuses and was left sided in all but one. A parallel or abnormal orientation of the great arteries was seen in all fetuses. Only five of 34 cases were isolated from a cardiac viewpoint. There was a ventricular septal defect in 21, Ebstein's anomaly in 8, pulmonary stenosis in 12, coarctation of the aorta in 6, tricuspid atresia in 2, congenital heart block in 2 fetuses prenatally, and dextrocardia in 6. Nineteen babies survived; one baby died in the neonatal period and three in infancy. Eleven pregnancies were interrupted.
Conclusions: A left sided or posterior right ventricle, identified by reversed differential insertion or the moderator band, in association with abnormal orientation of the great arteries should alert a sonographer to the diagnosis of double discordance. Isolated cases are rare prenatally (14%) but the short term survival in our fetal series is good. Where pregnancy was continued, 19 of 23 babies (82%) remained alive.

Atrioventricular and ventriculoarterial discordance, also known as congenitally corrected transposition of the great arteries (CCTGA), is a complex type of heart lesion that accounts for less than 1% of infants with congenital heart disease.[1–3] One of the original descriptions of CCTGA was by Karl von Rokitansky in 1875,[4] who described the hearts of a 4 month old girl and an 11 month old boy. In these hearts the great arteries were transposed, but the ventricles and the ventricular septum were arranged such that the transposition was physiologically corrected by allowing venous blood to enter the pulmonary artery and arterial blood to enter the aorta.

This type of cardiac malformation encompasses a very heterogeneous group of abnormalities, but the characteristic feature of all cases is discordance at both the atrioventricular and ventriculoarterial junctions. This double discordance results in a physiologically corrected circulation, but the left ventricle supports the pulmonary circulation and the right ventricle supports the systemic circulation. The goal of this study was to highlight features of CCTGA that should be recognisable during fetal heart scanning to improve the prenatal detection of this condition. In addition, the spectrum of this abnormality seen during fetal life has been documented and the outcome associated with both the isolated cases and cases occurring with other cardiac malformations has been evaluated.

METHODS

This study was a retrospective analysis and audit of data and echocardiograms already performed for clinical reasons, so ethical approval was not necessary.

Patient population

The study population was selected from patients referred to a tertiary centre for fetal cardiology between January 1993 and December 2003. During this period, 15 861 pregnant women at high risk of having a baby with congenital heart disease were scanned in our unit and structural congenital heart disease was diagnosed prenatally in 1913 fetuses. In 34 fetuses (1.8%) seen sequentially, CCTGA was diagnosed prenatally and these form the study group. The majority of fetuses, 31 of 34 (91%), were referred because of a suspected cardiac abnormality noted during an obstetric anomaly scan. One fetus was referred because of a family history of congenital heart disease, one because of maternal diabetes, and one because of an increased nuchal translucency measurement. The median gestational age at presentation was 20 weeks (range 15–31 weeks). In 28 of 34 fetuses (82%) the diagnosis was confirmed either postnatally or, where pregnancy was interrupted, at post mortem examination. However, for six fetuses, parental permission for necropsy was refused. During the study period a complex cardiac abnormality was diagnosed prenatally in two fetuses who

Table 1 Gestational age, referral reason, cardiothoracic ratio (CTR), and cardiac position in 34 fetuses

Patient	Gestational age (weeks)	Referral reason	CTR	Heart angle (in degrees left or right midline)
1	22	?CHD	0.49	40° left
2	23	?CHD	0.47	53° left
3	19	?CHD	0.49	25° right
4	19	?CHD	0.4	60° left
5	20	?CHD	0.49	40° right
6	17	?CHD	0.49	0
7	20	?CHD	0.5	55° left
8	22	?CHD	0.46	40° left
9	19	?CHD	0.49	50° left
10	21	?CHD	0.48	45° right
11	20	?CHD	0.51	50° left
12	18	?CHD	0.44	30° right
13	21	?CHD	0.45	60° left
14	17	?CHD	0.48	40° right
15	22	?CHD	0.51	24° left
16	26	?CHD	0.53	50° left
17	31	?CHD	0.6	55° left
18	20	?CHD	0.58	42° left
19	24	?CHD	0.58	40° left
20	20	?CHD	0.45	36° right
21	23	?CHD	0.45	65° left
22	15	FH	0.51	60° left
23	18	?CHD	0.51	35° left
24	20	?CHD	0.49	50° left
25	21	?CHD	0.43	50° left
26	21	?CHD	0.44	60° left
27	21	?CHD	0.47	40° left
28	20	?CHD	0.47	30° left
29	20	?CHD	0.70	50° left
30	21	?CHD	0.62	50° left
31	22	?CHD	0.51	48° left
32	20	Diabetes	0.62	48° left
33	22	?CHD	0.55	50° left
34	15	Nuchal oedema	0.5	36° left

?CHD, suspected congenital heart disease during obstetric ultrasound scanning; FH, family history.

Table 2 Echocardiographic features identified

Patient	Differential insertion	Moderator band seen	First vessel from 4 chambers	Parallel or abnormal orientation of great arteries
1	Reversed	Yes	PA	Yes
2	Reversed	Yes	PA	Yes
3	Reversed	Yes	PA	Yes
4	Reversed	Yes	PA	Yes
5	Reversed	Yes	PA	Yes
6	Not seen	Yes	PA	Yes
7	Reversed	Yes	PA	Yes
8	Reversed	Yes	PA	Yes
9	Reversed	Yes	PA	Yes
10	Reversed	Yes	PA, but difficult	Yes
11	Reversed	Yes	PA	Yes
12	Reversed	Yes	PA	Yes
13	Not seen	Yes	PA	Yes
14	Reversed	Yes	PA	Yes
15	Reversed	Yes	PA	Yes
16	Reversed	Yes	PA	Yes
17	Reversed	Yes	PA	Yes
18	Reversed	Yes	PA	Yes
19	Reversed	Yes	PA	Yes
20	Not seen	Yes	PA, but difficult	Yes
21	Not seen	Yes	PA	Yes
22	Reversed	Yes	PA	Yes
23	Reversed	Yes	PA	Yes
24	Reversed	No	PA	Yes
25	AV valve atresia	No, RV hypoplasia	PA	Yes
26	Not seen	Yes	PA	Yes
27	Reversed	Yes	PA	Yes
28	AV valve atresia	No, RV hypoplasia	PA	Yes
29	Reversed	No	PA	Yes
30	Reversed	Yes	PA, but difficult	Yes
31	Not seen	Yes	PA	Yes
32	Reversed	Yes	PA	Yes
33	Reversed	Yes	PA	Yes
34	Reversed	Yes	PA	Yes

AV, atrioventricular; PA, pulmonary artery; RV, right ventricle.

had CCTGA diagnosed as part of the complex abnormality postnatally. These two fetuses were excluded from the study.

Echocardiographic features

The fetal echocardiograms were recorded by a Toshiba SSA 270A ultrasound system (Toshiba Medical Systems, Crawley, UK) and Hewlett Packard 1000, 2000, and 5500 ultrasound systems (Philips Inc, Andover, Massachusetts, USA). The videotape recordings of all fetal echocardiograms were analysed retrospectively to document the features of CCTGA that were identifiable on the scan images and that had helped to make the original diagnosis. These features were sought in both the four chamber and great artery views. In the four chamber view this included reversed differential insertion of the atrioventricular valves and identification of the moderator band of the right ventricle to help recognition of this chamber. The atrioventricular valves are differentially inserted because the setting of the atrioventricular valves is more apical in the right ventricle. The phrase ''reversed differential insertion'' is used here to indicate that the more apically positioned atrioventricular valve, although still a marker of the right ventricle, is not in the correct position. The origin and orientation of the great arteries were also documented.

The cardiothoracic ratio was measured retrospectively by on-screen calibration of a still frame of the video record. The angle between the cardiac axis and the midline was also measured to assess the cardiac position. The cardiothoracic ratio usually increases slightly with advancing gestation, with the 95th centile increasing from 0.5 at 18 weeks to 0.55 at term.[5] The normal axis lies at a 45° angle (range 22–75°) to the left of an anteroposterior line drawn from the spine to the anterior chest wall.[6]

The situs had been determined at the time of the original scan by establishing left and right in the fetus, noting the position of the heart and the arrangement of the aorta and inferior vena cava in the abdomen.

Other abnormalities

Additional cardiac abnormalities and any associated extracardiac abnormalities were documented.

Outcome

The outcome data were derived from our extensive departmental database supplemented with information from postnatal echocardiograms and retrospective examination of postmortem reports.

Statistical analysis

The statistical analysis in this study was limited to descriptive statistics.

RESULTS

Tables 1–4 summarise the results.

Table 3 Associated cardiac lesions

Patient	Isolated	VSD	PS	PAt	EA	TR	Coarct	CHB	Other
1	No	No	No	No	Yes	Yes	No	No	None
2	No	Yes	No	No	Yes	Yes	Yes	No	None
3	Yes	No	No	No	No	No	No	No	None
4	No	Yes	No	No	No	No	Yes	No	None
5	No	Yes	Yes	No	No	No	No	No	None
6	No	Yes	Yes	No	No	No	No	Prenatal	None
7	No	Yes	No	No	No	No	No	No	None
8	Yes	No	No	No	No	No	No	No	None
9	Yes	No	No	No	No	No	No	No	None
10	No	Yes	Yes	No	No	No	No	No	None
11	No	Yes	No	No	No	No	No	Postnatal	None
12	No	Yes	Yes	Postnatal	No	No	No	No	None
13	No	No	Yes	No	No	No	No	Postnatal 2:1 block	None
14	No	Yes	Yes	No	No	No	No	No	None
15	No	Yes	Yes	No	No	No	No	No	None
16	No	Yes	Mild postnatal	No	No	No	No	No	None
17	No	No	Yes	No	No	No	No	No	None
18	Yes	No	Mild postnatal	No	No	No	No	No	None
19	No	Yes	No	No	No	No	No	No	None
20	No	Yes	Yes	No	No	No	No	No	None
21	No	Yes	No	No	No	No	No	No	None
22	No	No	No	No	Yes	No	No	Prenatal	None
23	Yes	No	No	No	No	No	No	No	None
24	No	No	No	No	Yes	No	No	No	None
25	No	Yes	No	Yes	No	No	No	No	AV valve atresia
26	No	Yes	Yes	No	No	No	No	No	None
27	No	Yes	Yes	Yes	No	No	No	No	None
28	No	Yes	No	No	No	No	Yes	No	AV valve atresia
29	No	No	No	No	Yes	Yes	No	No	None
30	No	Yes	No	No	Yes	Yes	No	No	None
31	No	Yes	Yes	No	No	No	No	No	None
32	No	Yes	No	No	Yes	No	No	No	None
33	No	No	No	No	No	No	Yes	No	None
34	No	No	No	No	Yes	Yes	No	No	None

CHB, complete heart block; Coarct, coarctation of the aorta; EA, Ebstein's anomaly; PAt, pulmonary atresia; PS, pulmonary stenosis; TR, tricuspid regurgitation; VSD, ventricular septal defect.

Echocardiographic features

Reversed differential insertion

Reversed differential insertion of the atrioventricular valves was easily identified in 26 of 34 fetuses (76%) (fig 1). Of the fetuses where reversed differential insertion could not be seen, five had an associated ventricular septal defect, in two there was associated atrioventricular valve atresia, and in the remainder the image quality was poor.

Recognition of right ventricle by moderator band

The right ventricle could be identified by the moderator band (fig 2) in 30 of 34 fetuses (88%) and was left sided in all but one (case 12), where it was right sided in association with complete situs inversus.

Origin of the great arteries

A parallel or abnormal orientation of the great arteries was seen in all fetuses. The pulmonary artery was the first vessel seen moving cranially from four chambers (figs 3 and 4). The aorta was anterior and to the left in all fetuses with the exception of the one fetus with situs inversus.

Combination of features

All the above features could be identified in 24 of 34 fetuses (71%).

Cardiac position and size

The cardiothoracic ratio was increased in four fetuses, of whom three had associated Ebstein's anomaly. There was mediastinal shift towards the right in seven fetuses. Six of them had dextrocardia, one of whom also had complete situs inversus (case 12). In the seventh fetus the heart was in the midline (case 6).

Associated cardiac abnormalities

Table 3 shows the associated cardiac abnormalities, if any, in each fetus.

Isolated cases

Isolated CCTGA was present in only five of 34 fetuses (14.7%).

Ventricular septal defect

A ventricular septal defect was present in 21 fetuses (62%), of whom two had associated left sided atrioventricular valve atresia (tricuspid atresia).

Pulmonary stenosis

Twelve fetuses were thought to have pulmonary stenosis on the antenatal scan. This was confirmed in nine fetuses, one fetus had pulmonary atresia at postmortem, and in two fetuses no postmortem had been performed for confirmation. In a further two fetuses, pulmonary stenosis had not been suspected on the antenatal scan but mild pulmonary stenosis was documented postnatally.

Ebstein's anomaly

Ebstein's anomaly was diagnosed prenatally in eight babies (fig 5) and this was confirmed in five, with permission for postmortem being refused for three.

Coarctation of the aorta

An associated coarctation of the aorta was diagnosed in six fetuses prenatally (fig 4). This was confirmed in four babies: two postnatally, both of whom had a coarctation repair, and two at postmortem examination. For the other two, permission for postmortem was declined.

Table 4 Outcome and confirmation of diagnosis

Patient	Prenatal diagnosis	Outcome	Postnatal or PM diagnosis
1	CCTGA, EA	Alive	CCTGA, EA
2	CCTGA, VSD, EA, Coarct	Alive	CCTGA, VSD EA, Coarct
3	CCTGA	Alive	CCTGA
4	CCTGA, VSD, Coarct	Alive	CCTGA, VSD, Coarct
5	CCTGA, VSD, PS	Alive	CCTGA, VSD, Coarct
6	CCTGA, VSD, CHB PS	Alive	CCTGA, VSD, CHB, late diagnosis PS
7	CCTGA, VSD	Alive	CCTGA, VSD
8	CCTGA	Alive	CCTGA
9	CCTGA	Alive	CCTGA
10	CCTGA, VSD, PS	Alive	CCTGA, VSD, PS
11	CCTGA, VSD	Alive	CCTGA, VSD, CHB
12	CCTGA, VSD, PS	Alive	CCTGA, VSD PAt
13	CCTGA, PS	Alive	CCTGA, PS, 2:1 AV block
14	CCTGA, VSD, PS	Alive	CCTGA, VSD PS
15	CCTGA, VSD, PS	Alive	CCTGA VSD PS
16	CCTGA, VSD	Alive	CCTGA, VSD PS
17	CCTGA, PS	Alive	CCTGA, PS
18	CCTGA	Alive	CCTGA, mild PS
19	CCTGA, VSD, hypoplastic RV	Alive	AV discord, DORV VSD, straddling RAVV
20	CCTGA, VSD, PS	INFD	CCTGA, VSD PS
21	CCTGA, VSD	INFD	CCTGA, VSD
22	CCTGA, EA, CHB	INFD	CCTGA, EA, CHB
23	CCTGA	NND	CCTGA
24	CCTGA, EA	TOP	CCTGA, EA
25	CCTGA, TAt, VSD, Coarct	TOP	CCTGA, TAt, VSD Coarct
26	CCTGA, VSD, PS	TOP	CCTGA, VSD PS/PAt
27	CCTGA, VSD, PS	TOP	Refused PM
28	CCTGA, TAt, VSD, Coarct	TOP	Refused PM
29	CCTGA, EA, Coarct	TOP	Refused PM
30	CCTGA, VSD, EA	TOP	Refused PM
31	CCTGA, VSD, PS	TOP	Refused PM
32	CCTGA, VSD, EA	TOP	AV disc, DORV, VSD EA
33	CCTGA, Coarct, hypoplastic RV	TOP	CCTGA, Coarct, hypoplastic RV
34	CCTGA, EA	TOP	Refused PM

CCTGA, congenitally corrected transposition of the great arteries; DORV, double outlet right ventricle; INFD, infant death; NND, neonatal death; PM, postmortem; RAVV, right atrioventricular valve; TAt, tricuspid atresia; TOP, termination of pregnancy.

Tricuspid regurgitation

Tricuspid regurgitation was noted in five fetuses on the prenatal scan, all of whom had associated Ebstein's anomaly.

Heart block

Two babies had associated complete heart block during fetal life and one developed this postnatally. A further baby developed 2:1 atrioventricular block postnatally.

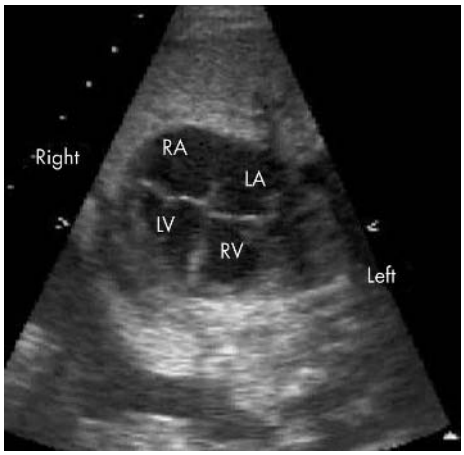

Figure 1 Four chamber view of the heart illustrating reversed differential insertion. The left sided atrioventricular valve is more apically positioned and thus the right ventricle (RV) is left sided and connected to the left atrium (LA). LV, left ventricle; RA, right atrium.

Extracardiac abnormalities

One baby had cataracts diagnosed postnatally and died at the time of cataract surgery. No identified chromosomal abnormalities were identified in any of the babies in this study.

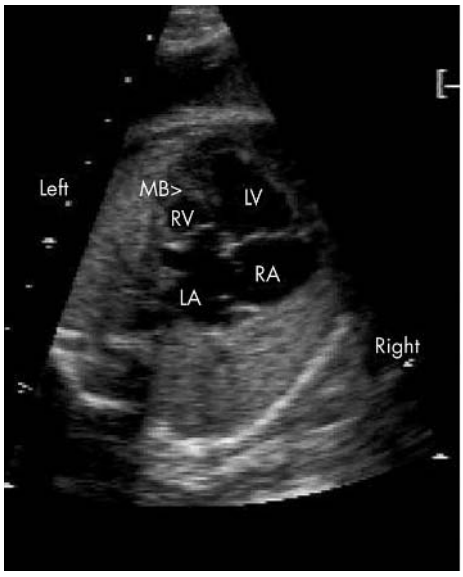

Figure 2 In this example, the right ventricle (RV) is also left sided but appears smaller than the left ventricle (LV). The moderator band (MB) and reversed differential insertion are both easily recognised.

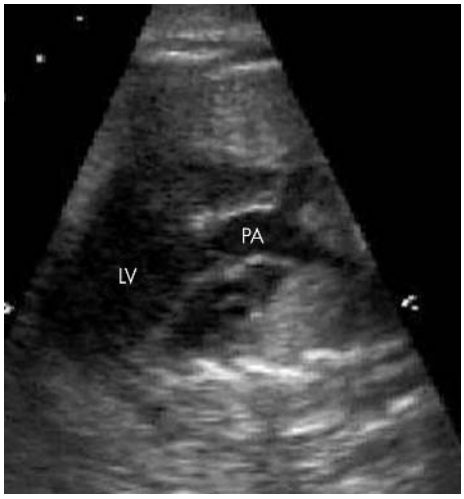

Figure 3 Moving cranially from the four chamber view seen in fig 1, the first vessel visualised is the pulmonary artery (PA).

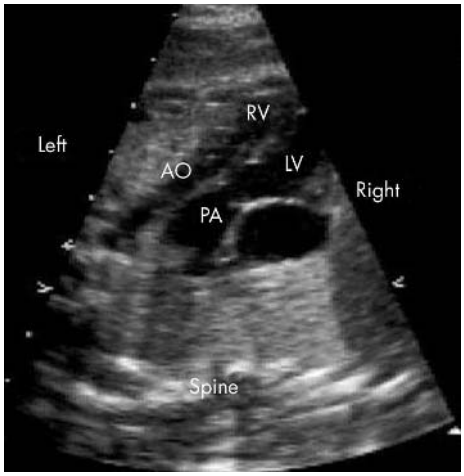

Figure 4 View of the great arteries of the example shown in fig 2. The great arteries arise in a parallel orientation and the aorta (AO) is anterior and to the left of the pulmonary artery (PA). The aorta is significantly smaller than the PA due to an associated coarctation of the aorta.

Outcome

Table 4 shows the outcome of the 34 fetuses. This table also allows comparison of the prenatal and postnatal diagnoses. There are 19 survivors, one baby died in the neonatal period, and three died in infancy. Parents elected to terminate 11 pregnancies (32%). However, for six of these fetuses the parents refused permission for necropsy so that the diagnosis could not be confirmed.

DISCUSSION

This diverse category of cardiac abnormality can be diagnosed with a high degree of accuracy in specialised centres, but may be confusing for sonographers and those inexperienced at examining the fetal heart. Thus, CCTGA may not easily be identified during routine obstetric ultrasound investigation, unless there are associated malformations such as Ebstein's anomaly or an absent atrioventricular connection. The fetal spectrum is thus biased towards these cases. However, prenatal detection during obstetric screening is clearly possible, as the majority of fetuses (91%) in this series were referred because of a suspected problem during routine obstetric screening.

The following echocardiographic features should alert the sonographer to this condition.

Firstly is *reversed differential insertion of the atrioventricular valves*. This was identified echocardiographically in 76% of our fetuses. Although expected in this condition, it is not consistently seen, particularly if there are associated lesions, such as a ventricular septal defect, or if image quality is suboptimal. In this situation the moderator band may be more helpful to identify the right ventricle.

Secondly is *the moderator band in the left sided or posterior ventricle*. In 88% of our fetuses the moderator band was visualised and helped to identify the right ventricle.

Thirdly is *abnormal orientation of the great arteries*. In the normal heart the first vessel viewed moving cranially from four chambers is the aorta. Loss of the normal crossover of the great arteries and the first vessel viewed when moving cranially from the four chamber view being the pulmonary artery should alert the sonographer. However, this finding may also be seen in ventriculoarterial discordance with atrioventricular concordance. The first two features also need to be sought to make a correct diagnosis.

All the above features were identified in 71% of fetuses in this study.

Isolated cases of CCTGA without other cardiac malformations are rare. In one postnatal series only 9% of CCTGAs were isolated.[7] In our series five of 34 (14.7%) were isolated. The most commonly seen associated malformations are ventricular septal defects, pulmonary stenosis or atresia, and tricuspid valve malformations.[1 8–11] Conduction abnormalities are also common postnatally in association with CCTGA.[1 10 12 13] Prenatally the diagnosis of CCTGA and conduction disturbances is much less common, although this has been reported.[14 15]

The natural history and clinical presentation are variable and depend on the presence and severity of associated cardiac lesions. Long term survival in this condition is possible, particularly if there are no associated abnormalities.[16 17] Huhta *et al*[18] described the overall survival of 70% at five years and 64% at 10 years. More recently, Rutledge *et al*[19] reported a survival of 92% at five years, 91% at 10 years, and 75% at 20 years. In this latter study, as in others, risk factors for mortality were identified as tricuspid regurgitation, right ventricular dysfunction, and complete atrioventricular block. In our fetal series, the patients are still relatively young and we cannot comment on long term survival, as regurgitation

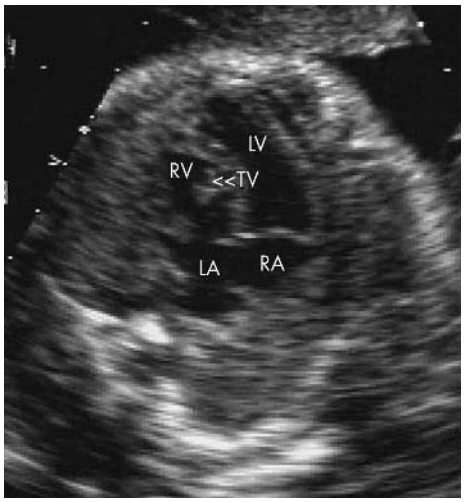

Figure 5 Example of atrioventricular discordance associated with Ebstein's anomaly of the tricuspid valve (TV).

of the systemic atrioventricular valve, right ventricular dysfunction, and atrioventricular block can all be progressive.[19 20] However, in the shorter term, only five babies in our series were noted to have tricuspid regurgitation prenatally, all of whom had associated Ebstein's anomaly. Of these, three pregnancies were terminated and two survived. A further 17 babies are still alive, none of whom had tricuspid regurgitation on the prenatal echocardiogram. Congenital heart block was present at diagnosis or developed in three babies, of whom two are alive and one died in infancy.

The ability of the right ventricle to sustain the systemic circulation over time is of major concern in the long term for patients with CCTGA. Conventional repair leaves the right ventricle as the systemic ventricle but patients undergoing a biventricular repair are more likely to develop right ventricular dysfunction than are patients whose symptoms are palliated or who do not undergo surgery.[19 21] Right ventricular function can also deteriorate with time.[20] Thus, the double switch procedure has been advocated as a treatment option for this group of patients, as this restores the morphological left ventricle as the systemic ventricle. However, although favourable early and midterm results have been reported for this procedure, long term follow up is still needed to evaluate the overall benefits of this option.[21–24] None of the patients in this fetal series have progressed to having a double switch and at present either are receiving palliative treatment or have not undergone surgery.

Conclusions

CCTGA is a well defined and specific cardiac abnormality that may be associated with a heterogeneous spectrum of other cardiac abnormalities and that can be accurately identified prenatally. Specific echocardiographic features can be recognised by obstetric sonographers during obstetric ultrasound screening, which may improve the prenatal detection of this type of malformation. The short term outlook for babies with a prenatal diagnosis is good but longer term follow up is needed. With improving surgical techniques the longer term outlook may become more favourable and this will affect the counselling of parents prenatally.

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We declare that there are no competing interests.

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