Counseling and outcomes of antenatally diagnosed congenital heart anomalies in Turkey

Türkiye'de antenatal tanısı konmuş doğumsal kalp anomalilerinde danışmanlık ve sonuçları

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ABSTRACT

Objective: To determine the clinical outcomes and decisions of families of fetuses with prenatally-diagnosed cardiac abnormalities.
Methods: Prenatally diagnosed cases (n=155) with congenital heart disease were retrospectively categorized according to the Allan-Huggon grading system: Group A (cardiac disease associated with severe / lethal extracardiac disease); Group B1 (low risk with a postnatal prognosis); Group B2 (moderate risk, amenable to surgical repair with a low mortality); and Group B3 (high risk, associated with high mortality after surgery). Neonatal outcomes, including termination of pregnancy, were recorded for 18 months of follow-up after counseling the parents. Student's t-test, Mann-Whitney U, Pearson's Chi-square test and Fischer's exact Chi-square test were used for statistical analyses.
Results: One hundred forty-five cases completed follow up. Thirty-nine cases (Group A) were associated with extracardiac lethal defects and the pregnancies were terminated; these cases were excluded from statistical evaluation. Twenty parents in Group B3 opted also for termination. The survival rates of ongoing pregnancies after 18 months of follow-up between the three cardiac abnormality Groups (Group B1, n=37; Group B2, n=12; and Group B3, n=37) were 89.2%, 66.7%, and 13.5%, respectively. Significance was present between the survival rates of the three Groups [Group B3 vs. Group B1: p=0.0001; OR: 52.8 (12.9-214.5); Group B3 vs. Group B2: p=0.0009; OR: 12.8 (2.8-58.9); Group B2 vs. Group B1: p=0.087; OR: 4.12 (0.84-20.2)].
Conclusion: Our practice and the findings reported herein support the efficacy of this staging system and counseling parents of fetuses for congenital heart diseases. *(Anadolu Kardiyol Derg 2011; 2: 137-45)*
Key words: Congenital heart defects, counseling, outcome, extracardiac malformations, ultrasound, fetal echocardiography

ÖZET

Amaç: Prenatal tanı almış, fetal kalp anomalilerinin, klinik sonuçları ve aile kararlarının değerlendirilmesi.
Yöntemler: Prenatal doğumsal kalp anomali tanısı konmuş olgular (n=155), retrospektif yöntemle Allan ve Huggon'ın sınıflandırması doğrultusunda, Grup A (doğumsal kalp anomalisine eşlik eden ağır / ölümcül kalp dışı patoloji); Grup B1 (postnatal prognoz açısından düşük risk), Grup B2 (orta risk, düşük mortalite oranı ile uyumlu cerrahi mortalite) ve Grup B3 (yüksek risk-cerrahi sonrası için yüksek mortalite) olarak sınıflandırıldı. Ailelere verilen, danışma sonrası yenidoğan verileri, buna gebelik sonlandırması dahil edilmiştir ve 18 ay süreyle izlendi.
Bulgular: Olguların 145'inin izlemi tam olarak gerçekleştirilmiştir. Otuz dokuz olguda (Grup A) eşlik eden ölümcül kalp dışı anomaliler saptanmıştır ve bunların gebelik sonlandırılmasına karar verilmiştir; bu olgular istatistiksel değerlendirmeye alınmamıştır. Grup B3' teki olguların 20 tanesi de gebelik sonlandırmasına karar verdi. Diğer üç kalp anomalisi gruplarında, gebeliğin devamına karar veren ailelerde (Grup B1, n=37; Grup B2, n=12; Grup B3, n=37), 18 ayın sonunda sağkalım oranları sırasıyla %89.2, %66.7 ve %13.5 bulunmuştur. İstatistiksel verilerin değerlendirmesinde Student's t-test, Mann-Whitney U, Pearson's Ki-kare testi ve Fischer's exact Ki-kare testi uygulanmıştır. Sağkalım açısından 3 grupta istatistiksel anlamlılık saptanmıştır [Grup B3 ile Grup B1: p=0.0001; OR: 52.8 (12.9-214.5); Grup B3 ile Grup B2: p=0.0009; OR: 12.8 (2.8-58.9); Grup B2 ile Grup B1: p=0.087; OR: 4.12 (0.84-20.2)].
Sonuç: Uygulamalarımız ve istatistiksel sonuçlarımız, bu sınıflama sisteminin, doğumsal kalp anomalileri açısından ebeveynlere verilecek danışmada etkili olduğunu gösterdi. *(Anadolu Kardiyol Derg 2011; 2: 137-45)*
Anahtar kelimeler: Doğumsal kalp anomalileri, danışma, sonuçlar, kalp dışı anomaliler, ultrason, fetal ekokardiyografi

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Introduction

Congenital heart diseases (CHDs) are the most common congenital anomalies that affect 8 of 1000 live births and are the major causes of morbidity and mortality in the perinatal period (1-4). These malformations are 6.5 times more common than chromosomal abnormalities and 4 times more common than neural tube defects (5, 6). Furthermore, four of 1000 live births will be affected by severe congenital cardiac malformations, which account for 20% of neonatal deaths and up to 50% of infant deaths attributed to congenital anomalies (2, 6). Nevertheless differences exist in the rates of prenatal diagnosis and termination of CHDs across populations, and through time (7-9), which has an impact on the live birth prevalence of the more severe cases of CHDs, particularly for hypo-plastic left heart syndrome (HLHS) and related morbidity and mortality (3, 10). Counseling of parents has become increasingly essential, as the frequency of detecting of fetal abnormalities has increased. Such abnormalities may be identified by routine screening (11) or specifically sought in cases with a family history of an inherited or familial disorder.

Each abnormality is unique to the affected family and may arouse anxiety (12) or lead to long-term difficulties (13); however, the severity of anxiety depends on the seriousness and ease of correction. The role of the physician is to provide simplified information for a correct understanding and optimal decision-making. Effective counseling is therefore as important as establishing the correct diagnosis (14).

In the current study, we have examined the overall outcome of prenatally diagnosed fetal cardiac malformations and associated extracardiac abnormalities. We categorized clinical findings, according to the Allan-Huggon grading system, examined the decisions of families after appropriate counseling regarding defects and treatment options, and drew conclusions based on 18 months of postnatal follow-up of surviving neonates.

Methods

Patients

This was a retrospective observational study conducted between January 2004 and December 2007 involving 155 cases of 8953 pregnancies screened and diagnosed with CHDs in our perinatology unit.

Fetal echocardiography

All patients were scanned in our perinatology unit of İstanbul Bakırköy Maternity and Children Diseases Hospital by one of four experienced maternal-fetal medicine physicians-sonographers using a Voluson 730 Expert TM (multi-frequency convex transducer 2.0-7.0 MHz; GE Healthcare, Milwaukee, WI, USA).

When a heart defect was diagnosed or suspected, the physicians at the center performed a detailed anatomic assessment, including the fetal heart in the presence of a pediatric cardiologist. For optimal fetal heart screening, two-dimensional and Doppler imaging were performed in four-chamber, five-chamber, three-vessel, ductal arch, aortic arch positions, short-and long-axis views of great vessels. Pregnancies with visualization problems for fetal hearts were examined the following day at the latest. Cardiac *situs*, rhythm, venous inflow, atrial and ventricular chambers, atrioventricular and semilunar valves, ventriculo-arterial connections, aortic and the ductal arches were visualized in all cases.

Study protocol

The patients were divided into two groups. Group A had cardiac defects with associated severe extracardiac defects and as a result lead to a critical outcome, where termination of pregnancy (TOP) was offered. These cases were excluded from statistical evaluation. Group B had cardiac pathologies isolated or associated minor defects. Group B was also divided into three subgroups, as noted by Allan et al. (14) and modified with our clinical diagnostic results (Table 1). Group B1 had a good prognosis and was comprised of those easily treated with no affect on the child in the long-term. After counseling, we advised close follow-up for this group. Group B2 had an intermediate prognosis, and consisted of those defects that could be repaired surgically with low mortality, however long-term survival is to be affected. For this group, we advised a close follow-up and explained the risks to be confronted. Group B3 resulted in a bad prognosis and consisted of those cardiac lesions with a high mortality following surgery or required repeat surgery during childhood or likely to have cardiac compromise as a young adult. For group B3, we counseled parents concerning each pathology individually and a related prognosis was offered TOP.

Subsequent information was retrieved for all cases from our computerized database containing; gestational age at diagnosis, definition of cardiac defects, Doppler demonstration of flow defects, definition of extra-cardiac defects (if present), chromosomal anomalies (if available), pregnancy outcome and pediatric cardiac surgery (if present), and neonatal follow-up. Fetal karyotyping was offered in all cases. Karyotypes were available in 115 cases (74.2% of 155 cases). Karyotyping was primarily performed by fetal blood sampling, amniocentesis, and fluorescence in situ hybridization (FISH) analysis. When a karyotype was not obtained, it was considered to be probably normal. During the study period, we routinely checked for a 22q11 microdeletion only in truncal abnormalities.

After completion of the fetal anomaly work-up, including cardiac and extra-cardiac abnormalities, a multidisciplinary medical panel, including members of perinatology, pediatric cardiology, and related subdivisions of pediatrics and pediatric

Table 1. Definition of risk groups (modified from ref. 14)

Low risk CHD - Group B1 (little or no effect on life or lifespan)	**Moderate risk CHD - Group B2** (low mortality for surgery, but likely to affect long-term survival)	**High risk CHD - Group B3** (a high mortality for surgery or repeated surgeries likely during childhood or likely to be compromised cardiologically as young adults)
VSD	TOF	Ectopia cordis
Tricuspid regurgitation	Simple TGA	Common arterial trunk
(with normal karyotype)	Simple corrected TGA	TOF with pulmonary atresia
Situs inversus totalis	AVSD	Pulmonary atresia with IVS (some forms)
Ductus arteriosus aneurysm	Coarctation	Severe aortic stenosis
	DORV (some forms)	DORV
	Isolated TAPVR	DOLV
	Ebstein's anomaly without severe cardiomegaly	Complex TGA
	Pulmonary stenosis with VSD	Complex corrected TGA
		Tricuspid atresia, double inlet ventricle
		Tricuspid atresia with VSD with pulmonary stenosis
		Pulmonary atresia with IVS, mitral atresia
		HLHS, AVSD with DORV and right atrial isomerism
		AVSD with CHB and left atrial isomerism
		Ebstein's anomaly with severe cardiomegaly
		TAPVR with obstruction or with atrial isomerism syndrome
		Coronary Fistula with HLHS
		Dilated cardiomyopathy
		Interruption

AVSD - atrioventricular septal defect, CHB - complete heart block, DOLV - double outlet left ventricle, DORV - double outlet right ventricle, HLHS - hypoplastic left heart syndrome, IVS - intact ventricular septum, TAPVR - total anomalous pulmonary venous return, TGA - transposition of great arteries, TOF - tetralogy of Fallot, VSD - ventricular septal defect
Italic printed definitions are new added pathologies to proposed risk groups (14)

surgery, counseled the couple. Our hospital Ethical Committee and Perinatal - Neonatal Council offered patients an opportunity to discuss the prenatal findings, neonatal prognosis, and pregnancy management and options, including TOP. Only in cases with chromosomal abnormalities, cases with a postnatal lethal prognosis, and severe handicaps, as described in Groups A and B3, was TOP offered. The hospital Ethical Committee concurred with TOP after counseling and consent of the parents. TOP was performed due to feticide after 24 weeks gestation.

Turkish law (law no. 2827, paragraph 5; 27 May 1983) authorizes legal TOP in two distinct conditions: (a) voluntary TOP until 10 weeks in unwanted pregnancies and (b) elective TOP on medical grounds. Elective termination is possible at every stage of gestation with no stated upper gestation limit if there are serious maternal (ongoing pregnancy is life-threatening) or fetal (a high risk of severe disabilities or an untreatable fatal disease) circumstances. The legal process requires the agreement of one obstetrician and one associated physician who declares a maternal or fetal cause justifying elective TOP. Autopsy was offered to all patients after TOP was performed by our pathologists.

Follow-up

Neonatal cardiac echocardiography was always performed by the same pediatric cardiologist (KO) using a Siemens Acuson Cypress Cardiovascular System (pediatric cardiac 5.4-6.4 MHz wideband phased array transducer; Mountain View, CA, USA). All neonates that survived the 2nd day of life and all fetuses that died after birth due to associated cardiac, extra-cardiac, or chromosomal anomalies were examined. Prenatal and postnatal cardiac diagnoses were compared. All surviving neonates were followed up directly until 18 months of age. In the case of cardiac surgery, they were referred to three other cardiac surgery centers in İstanbul, but follow-up after surgery was continued in our center.

Ten of the 155 cases were lost during prenatal and / or postnatal follow-up. The study was closed to new follow-up data on 1 January 2008, but follow-up times for survivors have been calculated up to the last follow-up available rather than up to the closing date of the study.

Statistical analysis

A Statistical Package for Social Sciences (SPSS) for Windows, version 10.0 (Chicago, IL, USA) was used for the

analysis of data. In addition to descriptive statistics to compare quantitative variables, Student's t-test was used for normally distributed variables, and the Mann-Whitney U test was used for analysis of variables, which were not normally distributed. Groups were compared in 2x2 contingency tables using Pearson's Chi-square test or Fischer's exact Chi-square test. The results were evaluated at 95% confidence intervals and at a significance level of $p<0.05$.

Results

During the study period, 155 pregnancies (1.73% of 8953 cases) were evaluated. The final results of 145 cases were available, with 10 cases lost to follow-up during pregnancy and the postnatal period. The average age of pregnant women in the study was 27 years (maximum, 42 years; minimum, 17 years). At the time of diagnosis, the average gestational age was 26.5 weeks (maximum, 39 weeks; minimum, 14 weeks) and the median interquartile gestational age was 26 weeks (range, 21-32 weeks).

Group A, the associated extra-cardiac pathology group (Table 2), consisted of 39 cases with different lethal diagnoses or poor prognoses. All of the cases also had additional cardiac pathologies with more severe and serious extra-cardiac prognoses.

Group B1 (Table 3) consisted of 37 cardiac abnormalities, isolated and / or associated with mild extra-cardiac pathologies. Thirty-three cases (89.2% of 37 cases) survived after 18 months of neonatal follow-up. In this group, there were two intrauterine deaths and two neonates died in the postnatal period (one postpartum and one postoperatively, both with an additional prenatal diagnosis of cystic hygroma with a postnatal diagnosis of Noonan syndrome and Goldenhar syndrome).

Twelve cases formed Group B2 (Table 4), of whom 11 cases underwent surgery and eight survived after 18 months of follow-up, with a median survival rate of 66.7%.

The 57 cases in group B3 had severe cardiac pathologies and poor prognoses. Thirty-seven families made the decision to continue the current pregnancy (Table 5); the other 20 families opted for TOP (35.1% of 57 TOP cases; Table 6). Only five cases (13.5% of 37 cases) survived during follow-up. Of 37 cases, seven cases died in uterus, 14 cases died in the neonatal period without surgical intervention, and 11 cases died in the postoperative period. Cardiac and additional extra-cardiac abnormalities of these cases are shown in tables 5 and 6.

The outcomes of all 145 cases divided in 4 groups (Groups A, B1, B2, and B3) are presented in Table 7. Group A patients were excluded from statistical evaluation. Thirty-nine cases with severe associated extracardiac defects (Group A) and 20 cases in Group B3 opted for TOP. The outcome and statistics of 86 pregnancies from the different groups that opted to continue their pregnancies were calculated. There was a significant statistical difference between the three cardiac abnormality groups with respect to the parental decisions to continue pregnancy (Group B1, n=37; Group B2, n=12; and Group B3, n=57) in relation to cases in which the pregnancy continued (χ^2, 43.56; p=0.0001) (Chi-square=43.56; p=0.0001). Specific differences in life expectancy between all three groups existed (Group B3 vs. Group B1; p=0.0001; OR: 52.8 [12.9-214.5]; Group B3 vs. Group B2; p=0.0009; OR: 12.8 [2.8-58.9]; Group B2 vs. Group B1; p=0.086; OR: 4.12 [0.84-20.2]).

Prostaglandin E1 (PGE1) was not used and there were no neonatal intensive care unit (NİCU) admissions in the B1 group. In B2, two cases with transposition of great arteries and one case with coarctation received treatment with PGE1. The case with double outlet right ventricle and esophageal atresia was admitted to the NİCU until stabilization was achieved. Twenty cases in the B3 group were treated with PGE1 and 12 cases were admitted to the NİCU.

Of 86 pregnancies, postnatal echocardiography was possible in 77 cases because of intrauterine deaths in 9 cases. Sixty- two cases (80.5%) were in complete agreement. In four cases, spontaneous closure of isolated ventricular septal defect (VSD) was detected during pregnancy, which also increased in the postnatal follow-up period. Of 11 cases, pre- and postnatal heart findings were not in complete agreement. Five cases had an over diagnosis and six cases had an under diagnosis, but the discrepancies did not alter the subgroups or the outcome.

Discussion

The diagnosis of a lethal or non-lethal cardiac defect is no longer appropriate as almost all abnormalities may be surgically corrected, although some have a high risk of mortality. There is

Table 2. Terminated cases, because of associated severe / lethal extra-cardiac pathologies (Group A)

Pathological classification	n = 39	Clinical diagnosis (n)
Chromosomal pathologies	23	Trisomy 21 (n=13), trisomy 18 (n=8), trisomy 13 (n=2)
Genitourinary pathologies	6	Bilateral renal agenesis (n=2), multicystic dysplastic kidneys associated with oligo- / anhydramnios (n=4)
Central nervous system pathologies	7	Anencephaly (n=1), holoprosencephaly (n=2), hemivertebra (n=1), vermis agenesis (n=1), neural tube defect (n=2)
Other	3	Severe generalized hydrops / septated cystic hygroma (n=3)

Table 3. Prognosis of cases in Group B1 with decision to continue the pregnancy

Case	Diagnosis (Cardiac + extracardiac pathology)	Outcome
1	Isolated VSD	Spontaneous closure
2	Isolated VSD	Spontaneous closure
3	Isolated VSD in twins	Spontaneous closure
4	Isolated VSD, IUGR	In close follow-up
5	Isolated VSD, PPROM	Premature birth, spontaneous closure
6	VSD, cystic hygroma	Noonan syndrome; postpartum death
7	VSD, Situs inversus totalis	Spontaneous closure
8	VSD, radius agenesis, IUGR	Holt-Oram syndrome, in close follow-up
9	VSD, omphalocele	Spontaneous closure, operated because of omphalocele
10	Isolated VSD	Spontaneous closure
11	Isolated VSD, IUGR	Spontaneous closure
12	VSD, oligohydroamnios	Intrauterine death (25 gestational weeks)
13	VSD	Operated because of anal atresia, spontaneous closure
14	VSD, polyhydroamnios	In close follow-up, postnatal diagnosis with 18 months was VSD + aortic regurgitation; prepared for operation
15	VSD, cystic hygroma	Persisting VSD with 2 years; mental retarded
16	VSD, oligohydroamnios, IUGR	operated
17	VSD, GİS obstruction	Spontaneous closure of VSD, operated because of GİS obstruction
18	VSD, polyhydroamnios, IUGR	Spontaneous closure of VSD, operated because of GİS obstruction
19	Isolated VSD	Spontaneous closure
20	VSD, cystic hygroma	Goldenhar syndrome; postop death
21	VSD, pelvicaliectasis	In close follow-up
22	Isolated VSD	In close follow-up
23	VSD, GİS obstruction	Intrauterine death (32 gestational weeks)
24	VSD, omphalocele	In close follow-up, operated because of omphalocele
25	VSD	In close follow-up
26	VSD	In close follow-up
27	VSD, IUGR	In close follow-up
28	VSD	In close follow-up
29	Tricuspid regurgitation	In close follow-up
30	Situs inversus totalis	In close follow-up
31	Situs inversus totalis	In close follow-up
32	Tricuspid regurgitation	In close follow-up
33	Tricuspid regurgitation	In close follow-up
34	Ductus arteriosus aneurysm, arrhythmia	In close follow-up
35	VSD, pelvicaliectasis	In close follow-up
36	VSD, tricuspid regurgitation	In close follow-up
37	Situs inversus totalis	In close follow-up

GIS - gastrointestinal system, IUGR - intrauterine growth retardation, Postop - postoperative, PPROM - preterm premature rupture of membranes, VSD - ventricular septal defect

a large variety of CHDs, from defects that require no treatment, such as small VSDs, to defects that can only be treated with palliative surgery, such as hypo-plastic heart syndromes. Our practice and findings in this study confirm the hypothesis and the efficacy of this staging system in counseling parents of fetuses with CHDs diagnosed prenatally.

The details of counseling depend on an accurate cardiac diagnosis, association with extra-cardiac malformations, gestational age, natural history of the malformation in intrauterine life, and surgical options. Previous reports indicate a major impact of prenatal diagnosis of cardiac defects on pregnancy outcome, with general termination rates of affected fetuses varying

Table 4. Prognosis of cases in Group B2 with decision to continue the pregnancy

Case	Diagnosis (Cardiac + extra-cardiac)	Outcome
1	AVSD, TOF	Postpartum death
2	TOF	Postoperative death
3	AVSD, oligohydroamnios	Postoperative death
4	AVSD, GİS obstruction, hydronephrosis, polyhydroamnios, Trisomy 21	Postoperative death
5	VSD, pulmonary stenosis	Operated, alive
6	DORV, esophagus atresia	Operated for DORV and esophagus, alive
7	TOF, cranial ventriculomegaly, polyhydroamnios	Operated, alive
8	TGA	Operated, alive
9	AVSD, trisomy 21	Operated, alive
10	Coarctation	Operated, alive
11	TOF (in twin)	Operated, alive
12	TGA	Operated, alive

AVSD - atrioventricular septal defect, DORV - double outlet right ventricle, GIS - gastrointestinal system, IUGR - intrauterine growth retardation, PPROM - preterm premature rupture of membranes, TGA - transposition of great arteries, TOF - tetralogy of Fallot, VSD - ventricular septal defect

between 8% and 50% (15-17). Clearly, the termination rate is influenced by a huge number of factors, such as the severity of cardiac and associated extra-cardiac defects, legal guidelines, parental and community beliefs, counseling strategies, and institutional experience, which may explain some of the discrepancies in the reported number of terminations (11, 18). Our data are in agreement with the literature (termination rate, 40.7%), although we had a two-step strategy for TOP. The first-step constituted associated extra-cardiac abnormalities with the greatest impact (terminated cases, 66.1%). These malformations had a more severe prognosis than existing cardiac pathology and therefore were the basic indication for TOP. There was a chromosomal anomaly incidence of 21.7% in our series, which is comparable to former studies (19-21). Additional systemic malformations are persuasive criteria for parents in making decisions regarding termination.

Parents should be counseled regarding the quality of life, as well as morbidity, and the post-operative lifetime is a more critical concept to comprehend (22). The skill of talking to parents is in making them understand, regardless of their level of education. Questions, such as why a cardiac defect has occurred and whether or not it will occur again in following pregnancies, should be answered with current information (14, 23). The decision to terminate may be based on the perception of pain or distress that the parents consider their baby will experience as a result of the abnormality, experienced by the siblings, and themselves (24).

One of the weakest areas of our information were the long-term outcomes, with a limitation of only 18 months of postnatal follow-up and a one-ventricle repair in survivors in Group B3. Expectant parents are hoping for a normal way of life for their child. It is noteworthy that they realize the limits of our experiences in this regard. Many complex cardiac defects do not fit readily into precise diagnostic categories (14). In addition, many cardiac defects are not uniform and have co-existing cardiac pathologies with a broad variety in the severity of prognosis. The second step in this study was an advised categorization in three groups, depending on cardiac pathology and prenatal security of diagnosis. Neither patients from Group B1, nor patients from Group B2 opted for TOP after counseling. Our aim was not to counsel or advice termination in Group B3, but to make parents clear about the severity and poor outcome they are confronted with.

Table 1 is only proposed to be a rough guideline to aid the concept of counseling. Every clinic would present a similar list with few differences, depending on the experience and individual level of confidence. This list may constitute a starting point with three different levels for those clinics without their own data for pathologies detected prenatally and follow-up cases, especially for counseling in parents. The next step would be a complete list for every pathology and related outcome. The effects and results of cardiac pathologies depend not only on accurate prenatal diagnoses, but also on postnatal appropriate support for pathologic cases and pediatric cardiac surgery. Therefore, our opinion is to counsel patients with real local data and not to only use statistics and numbers of other experienced clinics and offer euphemisms (25).

Study limitations

There were some limitations in this study. We just evaluated prenatally detected and followed pregnancy cases throughout in this study, and did not include all neonates born with CHD. Another point in this study was referring cases for cardiac surgery to three different surgery centers. Although all cases were delivered in our hospital, we were in need to refer some cases, but postoperative follow-up was achieved in surviving children. Because of the same reason of not having a pediatric cardiac surgery department in our hospital, our council does not include a pediatric cardiac surgeon for counseling. We were able to follow our series for only 18 months, although we hope to give further information in a later series.

Conclusion

Unfortunately, regardless of developments in genetics and ultrasound techniques and their impact on medical practice, the prevention of fetal anomalies relies on prenatal diagnosis and

Table 5. Prognosis of cases in Group B3 with decision to continue the pregnancy

Case	Diagnosis (Cardiac + extra-cardiac)	Outcome
1	DORV + tricuspid atresia	Operated, alive
2	DORV + HLHS (mitral atresia)	Postoperative death
3	DORV + Coarctation, oligohydroamnios, IUGR	Postoperative death
4	AVSD + CHB + left atrial isomerism, pleural effusion	Postpartum death
5	AVSD + left atrial isomerism	Postoperative death
6	DORV + dextrocardia, single umbilical artery	Postpartum death
7	Interruption, multicystic kidney	Postpartum death
8	AVSD + atrial isomerism	Postoperative death
9	Common arterial trunk	Intrauterine death (36 gestational weeks)
10	HLHS + AVSD	Postpartum death
11	DORV + AVSD, PPROM	Postpartum death
12	Tricuspid atresia + DORV	Operated, waiting for second operation
13	HLHS + AVSD, IUGR	Postoperative death
14	HLHS (mitral atresia)	Postpartum death
15	DORV + pulmonary atresia, IUGR, polyhydroamnios	Postoperative death
16	DORV + HLHS	Postpartum death
17	Tricuspid atresia + HRHS	Operated, alive
18	DORV + AVSD	Postpartum death
19	Complex TGA	Postoperative death
20	Common arterial trunk	Postpartum death
21	DORV + umbilical vein abnormality	Postoperative death
22	Tricuspid atresia + DORV	Operated, alive
23	Complex TGA	Intrauterine death (33 gestational weeks)
24	Tricuspid atresia + VSD + PS (twin pregnancy)	Postpartum death
25	HLHS (mitral atresia)	Postoperative death
26	Pulmonary atresia with IVS	Intrauterine death (25 gestational weeks)
27	Pulmonary atresia with IVS	Intrauterine death (31 gestational weeks)
28	TAPVR + connective tissue disease	Postpartum death
29	AVSD + severe aortic stenosis	Intrauterine death (28 gestational weeks)
30	Pulmonary atresia with IVS, ascites	Intrauterine death (22 gestational weeks)
31	AVSD + atrial isomerism	Postoperative death
32	Pulmonary atresia with IVS	Intrauterine death (28 gestational weeks)
33	HLHS + severe aorta stenosis	Postpartum death
34	HLHS + severe aorta stenosis	Postpartum death
35	Common arterial trunk	Postoperative death
36	DORV + PS+ coarctation	Postpartum death
37	Tricuspid atresia + VSD + PS	Operated, alive

AVSD - atrioventricular septal defect, CHB - complete heart block, DORV - double outlet right ventricle, HLHS - hypoplastic left heart syndrome, HRHS - hypoplastic right heart syndrome, IUGR - intrauterine growth retardation, IVS - intact ventricular septum, PPROM - preterm premature rupture of membranes, PS - pulmonary stenosis, TAPVR - total anomalous pulmonary venous return, TGA - transposition of great arteries, VSD - ventricular septal defect

termination of severe affected pregnancies. Some parents, because of their religious or cultural background, will decline termination. The decision to terminate may be based on the perception of pain or distress that the parents consider their baby will experience as a result of the abnormality, the consequences of intervention, the pain and distress experienced by the siblings, and the pain and distress experienced by the parents (24). Our practice and findings confirm the efficacy of this staging system in counseling parents of fetuses with CHDs diagnosed prenatally.

Conflict of interest: None declared.

Table 6. Terminated cases because of cardiac pathology of Group B3

Case	Cardiac Pathology	Additional extra-cardiac pathology	Gestational week in termination
1	TOF + pulmonary atresia	None	22
2	DOLV + pulmonary stenosis	None	23
3	DORV + HLHS (mitral atresia)	None	27
4	AVSD + isomerism	None	26
5	DORV + HLHS (mitral atresia)	None	24
6	DORV + HLHS (mitral atresia)	None	23
7	Tricuspid atrezi + HRHS	Intrauterine growth retardation	23
8	Severe aorta stenosis + EF	46 XY q+ polymorphism	23
9	AVSD + isomerism	Cranial mild ventriculomegaly, polydactyly, single umbilical artery	21
10	Single ventricle + truncus arteriosus	None	25
11	Ectopia cordis	Cantrell syndrome, omphalocele	15
12	Pulmonary atresia + IVS	None	24
13	AVSD + isomerism	Hydrops	18
14	Single ventricle + isomerism	Abdominal situs inversus	24
15	Severe aorta stenosis + EF	None	21
16	Ectopia cordis	Cantrell syndrome	18
17	Ectopia cordis	None	17
18	Severe aorta stenosis + EF	Oligohydroamnios	21
19	TOF + pulmonary atresia	Posterior urethral valve	21
20	Dilated cardiomyopathy	Intrauterine growth retardation, hydrops	25

AVSD - atrioventricular canal, DOLV - double outlet left ventricle, DORV - double outlet right ventricle, EF - endocardial fibroelastosis, HLHS - hypoplastic left heart syndrome, HRHS hypoplastic right heart syndrome, IVS - intact ventricular septum, TOF - tetralogy of Fallot

Table 7. Prognosis of cases and subgroups [1] (n=145)

	Decision, n[1]	Live born, n	Alive, n[2]	Alive[3] n[2]/n[1], %	Postpartum exitus, n	Postop exitus, n	Intrauterine demise, n	Total exitus, n	Mortality[3] (%)
Group A Associated extracardiac pathology	TOP: 39	-	-	-	-	-	-	-	-
Group B1	n = 37 - Continue: 37 - TOP: -	35	33	89.2	1	1	2	4	10.8
Group B2	n = 12 - Continue: 12 - TOP: -	12	8 (All 8 cases operated)	66.7	1	3	-	4	33.3
Group B3	n = 57 - Continue: 37 - TOP: 20	30	5 (All 5 cases operated)	13.5	14	11	7	32	86.5

Data are presented as percentages
[1]n=145, ten cases were lost during follow-up. Distribution of 10 lost cases was one case in Group B1, 2 cases in Group B2, and 7 cases in Group B3,
[2]alive cases after 18 months follow up,
[3]relations to cases decided for ongoing pregnancy: Chi-square=43.56, p=0.0001
Group B3 vs. Group B1: p=0.0001, OR: 52.8 (12.9-214.5)
Group B3 vs. Group B2: p=0.0009, OR: 12.8 (2.8-58.9)
Group B2 vs. Group B1: p=0.087, OR: 4.12 (0.84-20.2)
TOP- termination of pregnancy

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