


Impact of interdisciplinary counselling for parental decision-making in cases of pregnancies with prenatally diagnosed CHD

Original Article

Cite this article: Schneider K, Bousi D, and Stressig R (2023) Impact of interdisciplinary counselling for parental decision-making in cases of pregnancies with prenatally diagnosed CHD. *Cardiology in the Young* 33: 1172–1176. doi: [10.1017/S104795112200213X](https://doi.org/10.1017/S104795112200213X)

Received: 24 February 2022
 Revised: 26 May 2022
 Accepted: 11 June 2022
 First published online: 2 August 2022

Author for correspondence: Katja Schneider, Department of Neonatology, GFO Kliniken Bonn, Robert-Koch-Straße 1, 53115 Bonn, Germany. Tel: +49 151 5694223. Fax: +49 228 5052903. E-mail: katja.schneider@gfo-kliniken-bonn.de

Katja Schneider¹ , Dafni Bousi² and Rüdiger Stressig³

¹Department of Neonatology, GFO Kliniken, Bonn, Germany; ²Department of Gynaecology and Obstetrics, Städtische Kliniken, Cologne, Germany and ³Department of Fetal Diagnostics, Pränatal Plus, Cologne, Germany

CHDs are the leading cause of congenital malformations, with a prevalence of 9 per 1000 births.¹ Prenatal screening and, thus, the prediction of foetal pathologies have steadily improved over the past 20 years.² In Germany, specialised prenatal diagnostics is based on a maternal risk profile, such as increased maternal age, diabetes, or multiple pregnancies. Due to the increased risk profile, the number of women examined is rising and, therefore, also the number of malformations detected prenatally.

Especially in the case of complex duct-dependent heart defects, prenatal detection and planning of a specific treatment have been demonstrated to improve the postnatal outcome of children by ensuring the births of affected infants in appropriate centres.^{3–6} However, neither population-based nor single-centre studies comparing prenatally and postnatally diagnosed CHDs have demonstrated a positive impact on 1-year survival in the prenatally diagnosed group.^{7–9} Additionally, the quality of the prenatally raised diagnosis must be considered critically. Bensemlali et al. described a discordance between prenatal and postnatal diagnosis in 29% of infants with a major impact on neonatal treatment in 7.7% of the babies. In autopsy findings of intrauterine foetal deaths and terminated pregnancies, 8.5% of the foetal autopsies had a different diagnosis and 14.3% had major differences.⁶ Considering that, with regard to the termination of a pregnancy, every decision is irreversible and no error is correctable, the impact of an exact prenatal diagnosis and professional counselling is exceptionally high.¹⁰

Consequently, prenatal counselling represents an essential component of the long-term processing of and coping with CHDs.^{11–14} The positive benefits of a prenatally established diagnosis are offset by the dangers of parents being placed in a decision-making conflict by prenatal knowledge of a cardiac defect and disclosure of the option of pregnancy termination. Therefore, clinicians are forgoing a pivotal relationship with the families, and the quality of that relationship and the memories will have lasting impact.¹⁵

The aim of this study is to evaluate the impact and long-term follow-up of interdisciplinary prenatal consultation and diagnostics on parental decision-making in children with complex CHDs. For this purpose, two questionnaires were developed in which affected parents could report their experiences regarding their educational counselling interview.

Material and methods

This study was designed as a retrospective monocentric cohort study.

A database analysis was used to select pregnant women whose foetuses were prenatally diagnosed with a severe CHD. The analysis detected 425 women who were examined and counselled between 2004 and 2018 in a specialised prenatal outpatient clinic with an affiliated paediatric heart centre. Of the 425 women, 400 (94.1%) had continued the pregnancy and 25 (5.9%) had terminated the pregnancy following diagnosis and counselling.

Only pregnancies with foetuses with CHDs requiring prompt therapy after birth were included. Therefore, foetuses with isolated ventricular and/or atrial septal defects, moderate valvular obstruction, or regurgitation were excluded. All parents received at least one detailed, multi-disciplinary diagnostic and prognostic consultation with a gynaecologist, paediatric cardiologist, and paediatric cardiothoracic surgeon.

Ethical approval was provided by the medical faculty of the Rheinische Friedrich-Wilhelms-Universität (351/18).

Questionnaire development

Two questionnaires were developed – one for women who carried the pregnancy to term (I) and the other for women who terminated the pregnancy (II).

Both questionnaires were tested in advance on 10 subjects each. The questionnaires were sent to the women by mail in December, 2018 with an enclosed prepaid return envelope to the medical practice. A letter of information about the aim of the study project was attached to the questionnaire. The women were asked to answer the questions together with their partner if the

Table 1. Fetal and maternal characteristics of terminated pregnancies

CHD	Additional malformations	Chromosomal disorders	Influence of counselling on decision-making	Sufficiently informed (subjective)	Would you decide like this again?
d-TGA	No	No	Important	Yes	Indifferent
d-TGA	Yes	No	Important	Yes	Yes
TOF	No	Yes	Important	Yes	Yes
TOF	No	No	Decisive	No	Yes
CAT	No	No	Partial	Yes	Not answered
HLHS	No	No	Important	Yes	Indifferent
Complex	No	No	Partial	Yes	Yes

d-TGA = dextro-Transposition of the great arteries; TOF = Tetralogy of fallot; CAT = Common arterial trunc; HLHS = Hypoplastic left heart syndrome.

partner was present at the counselling session. No information on the identity of the subjects was asked for, thereby ensuring the anonymity of all results. All survey data collected are based on the subjective statements of the parents interviewed. The inclusion of return responses was completed in March, 2019.

The questions were divided into three categories:

- Impact and perception of the counselling situation,
- Option of indicated termination of pregnancy, and
- Current health status of the child and quality of life.

All data collected, including diagnoses, are based on subjective statements of the parents interviewed.

Statistics

To analyse the end points, the absolute and relative frequencies of responses were first presented descriptively. For this and further analyses, response categories could be categorised, depending on the distribution of frequencies.

Additionally, individual groups with regard to different CHDs were compared with respect to their responses on the basis of absolute and relative frequencies.

Hypothesis testing was two-sided by Pearson's Chi-squared test, and a *p*-value less than .05 was considered significant.

Results

The response rates were 45% (178 of 400) for Questionnaire I and 28% (7 of 25) for Questionnaire II, with an overall response rate of 44% (185 of 425).

The cardiac abnormalities and other characteristics of the foetuses and responses of the women who underwent abortion are presented in Table 1. All seven women who opted for abortions reported that they were satisfied with the counselling they received.

Because of the high difference in the number of subjects, a direct comparison of the two groups was not possible. Therefore, the following statistical analysis focused on the responses of the women who had continued their pregnancy after diagnosis of a severe CHD of the foetus (Questionnaire I).

Isolated heart defects were present in 78% of the children, additional organ malformations existed in 10%, and 7% of the children had chromosomal aberrations.

Hypoplastic left heart syndrome was the most common heart defect, accounting for 40% of cases (72 of 178). The further distribution of the included heart defects was as follows: tetralogy of Fallot (23 of 178), transposition of great arteries (18 of 178),

pulmonary atresia with hypoplastic right ventricle (17 of 178), atrioventricular septal defect (16 of 178), aortic stenosis (12 of 178), common arterial trunk (3 of 178), and Ebstein's anomaly (2 of 178). A total of 8% of the children had a complex CHD (15 of 178).

In 80% of all cases, the prenatal and postnatal diagnosis and prognosis coincided, with 12% of children faring worse than expected after birth and 7% faring better. A total of 77% of children are doing well today either with or without continuous therapy. Only 7.3% of the children (13 of 178) live with a limited quality of life today, according to their parents' assessment, while 27 children (15%) have died since birth.

For 73% of the respondents, the examination and counselling had an important or decisive influence on their subsequent decision. A total of 95% of the parents said they felt the quality was good or very good. Joint consultation with paediatric cardiologists and gynaecologists was also rated positively by 87% of the respondents. Regarding prognosis and assessment, it was reported in one-quarter of the cases that there were different assessments among the different specialists. The assessment as well as the recommendation to terminate the pregnancy did not tend to differ between the professional groups. The possibility of an abortion was discussed with 37% of the respondents, and 22% of them said they found it outrageous or highly distressing. In retrospect, four women said they regretted not having been asked explicitly about the possibility of terminating the pregnancy. Of all respondents, one woman said that, in retrospect, she would have made a different decision regarding continuation of the pregnancy.

A comparison of maternal responses, differentiated by children with a hypoplastic left heart syndrome and by children with all other heart defects, is presented in Table 2. Here, it is noticeable that in the hypoplastic left heart syndrome cohort, termination of pregnancy was discussed significantly more often than with the other groups of CHDs. While the accuracy of prenatal and postnatal diagnosis in the hypoplastic left heart syndrome group did not differ significantly from that of the other heart defects, there were significant differences in the assessment and prognosis of paediatric cardiologists and prenatal diagnosticians.

A total of 94% of women said they would opt for prenatal diagnostics in a future pregnancy, while more than 60% replied that they would want to know about any abnormality. Prospectively, 7 of the 178 (4%) women would make a different decision regarding abortion in another pregnancy than they did in the past. A total of 11% (3 of 27) of the mothers whose children later died said they would terminate their pregnancy in case of a similar CHD in a future pregnancy.

Table 2. Comparison between parental perceptions: children with HLHS versus children with other CHD

	Total N = 178	Others* N = 106	HLHS N = 72	Others versus HLHS P
Satisfied with the quality of counselling	168/177 (95%)	101/105 (96%)	67/72 (73%)	0,26
Concordance of prenatal and postnatal diagnosis and prognosis	143/178 (80%)	89/106 (84%)	54/72 (75%)	0,14
Different assessment of Gynaecologists and Cardiologists/Surgeons	45/178 (25%)	18/105 (17%)	27/72 (38%)	0,003
Good quality of life today	137/177 (77%)	88/104 (85%)	49/72 (68%)	0,006
Important influence of counselling on decision-making	131/178 (73%)	79/106 (75%)	52/72 (72%)	0,55
Addressing the issue of termination of pregnancy	63/172 (37%)	30/104 (29%)	33/68 (49%)	0,013
“I would decide differently today”	1/155 (0,7%)	1/96 (1%)	0/59 (0%)	0,42

*All heart defects except HLHS.

When asked what advice they would give to other affected parents today, almost all respondents recommended that they obtain as much information as possible and, if necessary, seek a second opinion.

Discussion

The present survey captures a large cohort of pregnant women with children with complex CHDs who received interdisciplinary consultation. The vast majority of women found these consultations to be positive and helpful in the decision-making process. Women's satisfaction with the counselling was independent of the severity of their child's heart defect and the subsequent quality of life.

In a review of the importance of prenatal parental counselling in congenital malformations, Marokakis et al. reported that counselling helped reduce parental worries.¹⁶ In a qualitative interview study of affected parents, Harris et al. underlined the impact of facing and dealing with parental uncertainties and their coping strategies. Envisioning best-case and worst-case scenarios supported by precise medical information can help parents identify their individual paths. It is pointed out that palliative care planning, with its coping strategies, can be used comparatively to learn how to better deal with uncertainties.¹⁷ This approach is improved by Hancock et al., who emphasise the importance of early palliative care planning and counselling to reduce anxiety in parents of foetuses with single-ventricle CHDs.¹⁸ This is in contrast to interviews with affected couples by Bratt et al., where reduced postnatal parental coherence and satisfaction were observed after prenatal diagnosis of a CHD compared to those with a postnatal diagnosis.¹⁹

The multi-disciplinary approach is also crucial to parental feelings of having received a comprehensive consultation. By including experts of the different disciplines involved, parents receive advice with a high level of expertise and assessments from a variety of perspectives. This is essential for the acceptance and understanding of the individual cardiac defect and the following procedures. In the present study, the agreement regarding the assessment of prognosis between paediatric cardiologists and gynaecologists was very high at 75%. This kind of professional consistency conveys security and increases parental trust. Only parents of children with hypoplastic left heart syndrome reported more frequent differences in assessment between paediatric cardiologists and prenatal diagnosticians regarding the postnatal prognosis of their child compared to the overall cohort. However, there was neither a tendency in one

direction nor in the other. This result perhaps illustrates the general uncertainty and subjectivity in the assessment of the prognosis of children with this special and severe CHD. As also described in the literature, a major factor in a retrospective positive evaluation of prenatal counselling might be a high concordance of prenatal and postnatal diagnosis and, moreover, prenatal prognosis and postnatal clinical development.²⁰

In the cohort presented, there was an extremely low rate of pregnancy termination. Only 5.9% of the women surveyed had decided to terminate their pregnancy after receiving extended prenatal diagnosis and counselling. Abortion rates of approximately 30–50% have been described in the literature after prenatal diagnosis of a severe CHD.^{21–23} In a comparative study, Vincenti et al. found that whether the diagnosis of a CHD was made prenatal or postnatal had no effect on infant survival, but it did have a significant impact on termination rates.⁹ The influence of prenatal diagnosis on pregnancy termination is also demonstrated in the epidemiologic work of Lytzen, where there was an increase in medically indicated abortions from 0.6 to 39% for a severe CHD over the past 17 years in Denmark. Concurrently, the incidence of live-born children with a CHD decreased significantly.²²

According to many studies, the significant factors that influence the decision for or against abortion are diverse. In addition to the severity of the heart defect and gestational age, religious, ethical and personal values have a powerful influence on parental decision-making.^{23,24} In countries with restrictive abortion regulations, the principle of non-directive prenatal counselling based on free decision-making, therefore, cannot take place. By removing abortion as an option, prenatal diagnostics has a completely different impetus, serving mainly as a diagnostic purpose. Conversely, decision-making is reduced to postnatal therapy management – compassionate care versus intensive care.

In the survey presented here, only 37% of the respondents actively discussed the option of termination of pregnancy during their counselling interviews. Only four women stated afterwards that they would have preferred discussing this option during pregnancy. In contrast, of the patients who were confronted with the possibility of termination of pregnancy, 22% expressed that addressing this option had been threatening and upsetting to them. A study by Hilton-Kamm et al. assessed how sensitively the discussion of the termination of a pregnancy is perceived, finding that a high percentage of parents feel pressured regarding abortion.¹¹ In a survey of 841 affected parents, Hilton-Kamm highlighted the particular influence of the way in which information is conveyed on

the parents' processing and, ultimately, their decision-making.¹³ Educational interviews are subjectively shaped by the personal experiences of the consulting physician and, above all, the in-house results in treatment, as found by Kon et al. in a multi-centre study.²⁵ What is remarkable in the results of this study is that although women who were pregnant with fetuses having hypoplastic left heart syndrome were significantly more likely to discuss termination of their pregnancy, the average rate of termination did not increase in this group.

The fact that only 4% of the women would decide differently about continuing with a repeat pregnancy if their child had a complex CHD is certainly an expression of the positive postnatal outcome and care of the children at the heart centre. More than two-thirds of the respondents stated that their child had a good quality of life. Although the quality of life of children with hypoplastic left heart syndrome was described as significantly lower compared to that found with other heart defects, it was also remarkably high here at 68%. These results underlie the findings of Ernst et al., who emphasise that bio-psychological factors and parental stress can have a greater impact on their children's quality of life than the severity of the disease.²⁶

In contrast to this, 29% of the women interviewed who had terminated their pregnancy after prenatal diagnosis of a CHD stated that they doubted their decision in retrospect, although they were fully satisfied with the quality of the consultation. A correlation between termination of pregnancy and the severity of the heart defect or additional malformations was not apparent.

The presented study is limited by its non-standardised design and its low response rate. Since a significant number of women in the cohort did not take part in this study, a bias towards more satisfied subjects cannot be excluded.

To maintain anonymity, no personal data were collected for this sensitive subject area. Therefore, no further analyses were possible with regard to the influence of socio-demographic factors on the perception and evaluation of counselling.

Furthermore, the significantly lower rate of terminated pregnancies in the present cohort compared to the literature might be biased. Since the counselling took place at a large, renowned heart centre, a pre-selection of affected parents could be assumed in advance. The fact that multi-disciplinary counselling that includes cardiac surgeons can influence the termination rate of pregnancies, particularly in the case of univentricular heart defects, has recently been described.²⁷

Even if the small number of cases prohibits a representative statement, it can be postulated that the decision to terminate or continue the pregnancy after prenatal diagnosis of a CHD depends only partly on the severity of the expected clinical picture. It seems that personal values, individual perceptions of quality of life, and social and financial resources appear to have an important influence on decision-making. This emphasises the special importance and responsibility of prenatal counselling and demonstrates the need for multi-professional support of affected couples. In this context, medical and psychosocial counselling and care play an important role. Further studies, especially designed as multi-centre standardised questionnaires, are needed to evaluate the influence of prenatal counselling on the decision-making of parents after the diagnosis of a severe CHD. In addition to the individual values and biographies of the parents, the special educational behaviour of the physicians as well as the therapeutic success rate of the cooperating heart centre should be in focus as influencing factors to be investigated.

Summary

The results of this survey of affected women indicate that prenatal counselling is of significant importance for parents of children with a CHD. Considering the below-average rate of abortions, it can be assumed that the high level of reputation at the centre and the setting of the counselling had a significant influence on parental decision-making. Additionally, a high concordance between prenatal and postnatal diagnosis among experts from different specialties and a good quality of life of the affected children might be reflected in the high level of retrospective satisfaction with prenatal counselling among the parents who took part in this study.

Acknowledgement. None.

Financial support. This research received no specific grant from any funding agency, commercial or not-for-profit sectors.

Conflicts of interest. None.

Ethical standards. The authors assert that all procedures contributing to this work comply with ethical standards of the relevant national guidelines on human experimentation and with the Helsinki Declaration of 1975, as revised in 2008, and have been approved by the institutional committees (The study was approved by the Medical Ethics Committee of the University Hospital Bonn, Germany ((AZ 351/18). Local Ethics Committee of the University Hospital Bonn agreed on using data collection by the survey without written informed consent of the included patients. A letter of information was added to the survey.)

References

1. van der Linde D, Konings EE, Slager MA, et al. Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. *J Am Coll Cardiol* 2011; 58: 2241–7.
2. Lytzen R, Vejstrup N, Bjerre J, et al. The accuracy of prenatal diagnosis of major congenital heart disease is increasing. *J Obstet Gynaecol* 2020; 40: 308–315.
3. Holland BJ, Myers JA, Woods CR Jr. Prenatal diagnosis of critical congenital heart disease reduces risk of death from cardiovascular compromise prior to planned neonatal cardiac surgery: a meta-analysis. *Ultrasound Obstet Gynecol* 2015; 45: 631–8.
4. Tworetzky W, McElhinney DB, Reddy VM, Brook MM, Hanley FL, Silverman NH. Improved surgical outcome after fetal diagnosis of hypoplastic left heart syndrome. *Circulation* 2001; 103: 1269–73.
5. Mahle WT, Clancy RR, McGaurn SP, Goin JE, Clark BJ. Impact of prenatal diagnosis on survival and early neurologic morbidity in neonates with the hypoplastic left heart syndrome. *Pediatrics* 2001; 107: 1277–82.
6. Bensemlali M, Stirnemann J, Le Bidois J, et al. Discordances between prenatal and post-natal diagnoses of congenital heart diseases and impact on care strategies. *J Am Coll Cardiol* 2016; 68: 921–30.
7. Wright LK, Ehrlich A, Stauffer N, Samai C, Kogon B, Oster ME. Relation of prenatal diagnosis with one-year survival rate for infants with congenital heart disease. *Am J Cardiol* 2014; 113: 1041–4.
8. Khoshnood B, Lelong N, Houyel L, et al. EPICARD study group. impact of prenatal diagnosis on survival of newborns with four congenital heart defects: a prospective, population-based cohort study in France (the EPICARD Study). *BMJ Open* 2017; 7: e018285.
9. Vincenti M, Guillaumont S, Clarivet B, et al. Prognosis of severe congenital heart diseases: do we overestimate the impact of prenatal diagnosis? *Arch Cardiovasc Dis* 2019; 112: 261–269.
10. Silverman NH, Moon-Grady AJ. Discordant fetal and post-natal diagnosis: can we do better? *J Am Coll Cardiol* 2016; 68: 931–3.
11. Hilton-Kamm D, Chang RK, Sklansky M. Prenatal diagnosis of hypoplastic left heart syndrome: impact of counseling patterns on parental perceptions and decisions regarding termination of pregnancy. *Pediatr Cardiol* 2012; 33: 1402–10.

12. Lee CK. Prenatal counseling of fetal congenital heart disease. *Curr Treat Options Cardiovasc Med* 2017; 19: 5.
13. Hilton-Kamm D, Sklansky M, Chang RK. How not to tell parents about their child's new diagnosis of congenital heart disease: an internet survey of 841 parents. *Pediatr Cardiol* 2014; 35: 239–252.
14. Menahem S. Ethical questions arising from counselling in fetal complex congenital heart disease. *Monash Bioeth Rev* 2012; 30: 62–7.
15. Bertaud S, Lloyd DFA, Sharland G, Razavi R, Bluebond-Langner M. The impact of prenatal counselling on mothers of surviving children with hypoplastic left heart syndrome: A qualitative interview study.
16. Marokakis S, Kasparian NA, Kennedy SE. Prenatal counselling for congenital anomalies: a systematic review. *Prenat Diagn* 2016; 36: 662–71.
17. Harris KW, Brelsford KM, Kavanaugh-McHugh A, Clayton EW. Uncertainty of prenatally diagnosed congenital heart disease: a qualitative study. *JAMA Netw Open* 2020; 3: e204082.
18. Hancock HS, Pituch K, Uzark K, et al. A randomised trial of early palliative care for maternal stress in infants prenatally diagnosed with single-ventricle heart disease. *Cardiol Young* 2018; 28: 561–570.
19. Bratt EL, Järholm S, Ekman-Joelsson BM, et al. Parental reactions, distress, and sense of coherence after prenatal versus postnatal diagnosis of complex congenital heart disease. *Cardiol Young* 2019; 29: 1328–1334.
20. van Velzen CL, Clur SA, Rijlaarsdam ME, et al. Prenatal diagnosis of congenital heart defects: accuracy and discrepancies in a multicenter cohort. *Ultrasound Obstet Gynecol* 2016; 47: 616–22.
21. Baardman ME, du Marchie Sarvaas GJ, de Walle HE, et al. Impact of introduction of 20-week ultrasound scan on prevalence and fetal and neonatal outcomes in cases of selected severe congenital heart defects in The Netherlands. *Ultrasound Obstet Gynecol* 2014; 44: 58–63.
22. Lytzen R, Vejlstrop N, Bjerre J, et al. Live-Born major congenital heart disease in Denmark: incidence, detection rate, and termination of pregnancy rate from 1996 to 2013. *JAMA Cardiol* 2018; 3: 829–837.
23. Gendler Y, Birk E, Tabak N, Koton S. Factors that influence parents' decision-making regarding termination of pregnancy after prenatal diagnosis of fetal congenital heart disease. *J Obstet Gynecol Neonatal Nurs* 2021; 50: 475–484.
24. Chenni N, Lacroze V, Pouet C, et al. Fetal heart disease and interruption of pregnancy: factors influencing the parental decision-making process. *Prenat Diagn* 2012; 32: 168–72.
25. Kon AA, Ackerson L, Lo B. How pediatricians counsel parents when no “best-choice” management exists: lessons to be learned from hypoplastic left heart syndrome. *Arch Pediatr Adolesc Med* 2004; 158: 436–41.
26. Ernst MM, Marino BS, Cassidy A, et al. Biopsychosocial predictors of quality of life outcomes in pediatric congenital heart disease. *Pediatr Cardiol* 2018; 39: 79–88.
27. Kim ST, Song J, Huh J, et al. The effect of multidisciplinary approach on the birth rate of fetuses with prenatally diagnosed congenital heart disease. *J Korean Med Sci* 2019; 34.