

# Fetal heart disease and coating strategies for the health team: An integrative review

Cardiopatía fetal e estratégias de enfrentamento para a equipe de saúde: Uma revisão integrativa

Cardiopatía fetal y estrategias de revestimiento para el equipo de salud: Una revisión integrativa

## RESUMO

Objetivo: Analisar as evidências científicas disponíveis na literatura acerca da cardiopatía fetal e estratégias de enfrentamento para a equipe de saúde. Método: Trata-se de uma revisão integrativa. Realizou-se a busca por artigos; com delimitação nos últimos 5 anos (2016-2021); nos idiomas português, inglês e espanhol; disponíveis na íntegra. Nas seguintes plataformas de dados: BDNF, DOAJ, LILACS, MEDLINE, SciELO, SCOPUS e Web of Science. Resultados: Os dados foram organizados e apresentados em figuras e tabelas. Dos 1509 estudos encontrados, 6 estava disponível na BDNF, 2 na DOAJ, 26 na LILACS, 890 na MEDLINE, 0 na SciELO, 33 na SCOPUS e 552 na Web of Science. Contudo, após a leitura permaneceram apenas os que atendiam aos critérios para inclusão e exclusão descritos na metodologia, 5 estudos. Conclusão: Este estudo pode auxiliar a disseminar a importância do diagnóstico precoce da cardiopatía congênita para que ocorra o acompanhamento e prestação da assistência adequada.

**DESCRIPTORIOS:** Cardiopatías Congénitas; Equipe de Assistência ao Paciente; Estratégias de Saúde.

## ABSTRACT

Objective: To analyze the scientific evidence available in the literature about fetal heart disease and coping strategies for the health team. Method: This is an integrative review. The search for articles was performed; with delimitation in the last 5 years (2016-2021); in the Portuguese, English and Spanish; available in full. On the following data platforms: BDNF, DOAJ, LILACS, MEDLINE, SciELO, SCOPUS and Web of Science. Results: The data were organized and presented in figures and tables. Of the 1509 studies found, 6 were available in BDNF, 2 in DOAJ, 26 in LILACS, 890 in MEDLINE, 0 in SciELO, 33 in SCOPUS and 552 in the Web of Science. However, after reading, only those that met the inclusion and exclusion criteria described in the methodology, 5 studies remained. Conclusion: This study may help to disseminate the importance of early diagnosis of congenital heart disease for the follow-up and provision of adequate care.

**DESCRIPTORS:** Heart Defects, Congenital; Patient Care Team; Health Strategies.

## RESUMEN

Objetivo: Analizar la evidencia científica disponible en la literatura sobre cardiopatía fetal y estrategias de afrontamiento para el equipo de salud. Método: Esta es una revisión integradora. Se realizó la búsqueda de artículos; con delimitación en los últimos 5 años (2016-2021); en portugués, inglés y español; disponible en su totalidad. En las siguientes plataformas de datos: BDNF, DOAJ, LILACS, MEDLINE, SciELO, SCOPUS y Web of Science. Resultados: Los datos fueron organizados y presentados en figuras y tablas. De los 1509 estudios encontrados, 6 estaban disponibles en BDNF, 2 en DOAJ, 26 en LILACS, 890 en MEDLINE, 0 en SciELO, 33 en SCOPUS y 552 en la Web of Science. Sin embargo, después de la lectura, solo aquellos que cumplieron con los criterios de inclusión y exclusión descritos en la metodología, permanecieron 5 estudios. Conclusión: Este estudio puede ayudar a difundir la importancia del diagnóstico precoz de la cardiopatía congénita para el seguimiento y la prestación de una atención adecuada.

**DESCRIPTORIOS:** Cardiopatías Congénitas; Grupo de Atención al Paciente; Estrategias de Salud.

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**INTRODUCTION**

**C**ongenital Heart Disease (CHD) is the most common cause of birth defects and a leading cause of still-birth. The CHD phenotype is often associated with other genetic abnormalities and syndromes. The incidence in live births is approximately 9.1/1,000. The prenatal diagnosis rate varies from 25.1% in certain series, and the highest diagnosis rate can reach up to 97%. CHDs are complex and include both genetic and non-genetic aspects. <sup>(1)</sup>

About 50 CHDs are severe enough to have symptoms in utero or immediately after birth, requiring specific treatment in the early hours or days of life. The prenatal knowledge of these anomalies greatly favors the clinical development of these babies, as it allows the programming of the ideal place of delivery, the gestational age and the appropriate method of delivery. <sup>(2)</sup>

Genetic factors that can lead to CHD include polygenic, chromosomal, genomic, and monogenic disorders. Indications for a detailed fetal cardiac examination, echocardiographic studies include fetal chromosomal abnormalities, fetal systemic edema, fetal heart rate abnormalities, single or multiple cardiovascular defects, and other known defects have a risk of heart failure. <sup>(3)</sup>

Infant mortality rates or severe neurodevelopmental disorders can be significantly increased when genetics is the underlying

cause of CHD. The American College of Obstetricians and Gynecologists has recommended the use of chromosomal microarray analysis (CMA) as a first-line genetic diagnostic method for fetuses with structural abnormalities on prenatal ultrasound. <sup>(4)</sup>

From studies that examined the application of CMA for prenatal or postnatal diagnosis, it was concluded that CMA increased the detection rate of pathogenic changes by 12.0% compared to karyotype analysis. This differs from other studies that focused only on the conditions necessary for the detection of chromosomal diseases in fetuses with CHD. However, prognostic information for fetuses undergoing prenatal diagnosis of CHD is limited. <sup>(5)</sup>

One of the main goals of prenatal diagnosis is the detection of severe CHD, whose prognosis most often depends on planning the delivery in a specialized referral center, although fetal echocardiography, traditionally indicated in high-risk pregnant women, is quite accurate, the majority of newborns with heart disease in all parts of the world are still born without diagnosis, as many cases of congenital heart disease occur in low-risk groups and are not detected by screening at the time of prenatal ultrasound. <sup>(1-5)</sup>

Since the emergence of fetal medicine as a specialty and recent advances in ultrasound, detection of fetuses with congenital malformations has become more common,

allowing earlier treatment and significantly reducing fetal and neonatal mortality. <sup>(2,4)</sup>

Considering the behavior of cardiologic mutations in the fetal phase, it is extremely important to know which of these mutations may end up evolving hemodynamically, still in the intrauterine environment, and which ones will need some type of conduct before the time of birth, whether through drug administration, intrauterine intervention or anticipating birth. <sup>(1,3)</sup>

Therefore, this study aims to analyze the scientific evidence available in the literature about fetal heart disease and coping strategies for the health team.

**METHOD**

This is a bibliographic, descriptive study of the integrative review type, with a qualitative approach. From the following steps: <sup>(1)</sup> elaboration of the guiding question and objective of the study; <sup>(2)</sup> definition of inclusion and exclusion criteria for scientific productions; <sup>(3)</sup> search for scientific studies in databases and virtual libraries; <sup>(4)</sup> analysis and categorization of the productions found; <sup>(5)</sup> results and discussion of findings. <sup>(6)</sup>

To raise the guiding question, the PICO strategy was used, a methodology that helps in the construction of a research question and search for evidence for a non-clinical research, where P = Population/Patient; I = Interest; and Co = Context (P: Fetal Heart Disease; I: Coping Strategies; Co: Multi-

disciplinary Care). Thus, the following guiding question was defined for the research: “What are the coping strategies presented by the health teams for fetal heart disease?”.

For the selection of articles, the following inclusion criteria were used: original article, available in full, published in the last 5 years (2016-2021) in Portuguese, English or Spanish, which responded to the objective of the study. Gray literature was excluded, as well as repeated publications of studies in more than one database and articles that did not answer the guiding question of the study and that allowed access through the Virtual Private Network (VPN) of the University of Pernambuco (UPE). The temporal delimitation in the last 5 years is justified in order to survey recent articles.

Data collection took place during January and February 2022 in the following Databases: Nursing Database (BDENF); Directory of Open Access Journals (DOAJ); Latin American and Caribbean Literature in Health Sciences (LILACS); Medical Literature Analysis and Retrieval System Online (MEDLINE); SCOPUS, Scientific Electronic Library Online (SciELO) and the Web of Science.

Articles indexed from the Health Sciences Descriptors (DeCS) were searched: “Cardiopatas Congênicas”, “Estratégias de Saúde”, “Equipe de Assistência ao Paciente”. The respective terms from the Medical Subject Headings (MeSH) were used: “Heart Defects, Congenital”, “Health Strategies”, “Patient Care Team”. The operationalization and the search strategy were based on the combination of the Boolean operator AND and OR, performing the search together and individually so that possible differences could be corrected (Chart 1).

The selection of studies was based on the Preferred Reporting Items for Systematic Review and Meta-Analyse (PRISMA) in order to assist in the development of articles.<sup>(7)</sup> At first, duplicate studies were eliminated by reading titles and abstracts. Of these pre-selected, a full reading was carried out in order to verify those that meet the guiding question and the inclusion/exclusion criteria. The final sample was then constructed with studies relevant to the pre-established

criteria (Figure 1).

After reading the selected articles, the studies were categorized, classifying the knowledge produced in levels of evidence according to Melnyk and Fineout-Overholt (8): level I, evidence is related to the systematic review or meta-analysis of randomized controlled clinical trials or from clinical guidelines based on systematic reviews of randomized controlled clinical trials; at level II, evidence derived from at least one well-designed randomized controlled clinical trial; at level III, evidence from well-designed clinical trials without randomization; at level IV, evidence from well-designed cohort and case-control studies; at level V, evidence from a systematic review of descriptive and qualitative studies; at level VI, evidence derived from a single descriptive or qualitative study; and at level VII, evi-

dence derived from the opinion of authorities and/or the report of expert committees.

A summary of the information in the corpus was obtained through an instrument: identification of the original article; authorship of the article; year of publication; parents; methodological characteristics of the study; and study sample. An analytical reading of the studies was carried out, identifying the key points for the hierarchy and synthesis of ideas.

Aiming at a better understanding and visualization of the main findings, the data were organized by presenting them in figures and tables, exposed in a descriptive way.

**RESULTS**

The studies surveyed are arranged showing their titles, authors, years of pu-

Chart 1 - Database search strategy. Recife, Pernambuco (PE), Brazil, 2022.

Databases	Search terms	Results	Selected
BDENF	(Heart Defects, Congenital AND Health Strategies) OR (Heart Defects, Congenital AND Patient Care Team)	6	0
DOAJ	Heart Defects, Congenital AND (Health Strategies OR Patient Care Team)	2	0
LILACS	(Heart Defects, Congenital AND Health Strategies) OR (Heart Defects, Congenital AND Patient Care Team)	26	0
MEDLINE	(Heart Defects, Congenital AND Health Strategies) OR (Heart Defects, Congenital AND Patient Care Team)	890	2
SciELO	Heart Defects, Congenital AND Health Strategies AND Patient Care Team	0	0
SCOPUS	Heart Defects, Congenital AND Health Strategies AND Patient Care Team	33	0
Web of Science	(Heart Defects, Congenital AND Health Strategies) OR (Heart Defects, Congenital AND Patient Care Team)	552	3
Total		1509	5

Source: Research data, 2022.

blication, levels of evidence, objectives and results. After reading the selected articles, the studies were categorized, classifying the knowledge produced on the topic, into levels of evidence, mostly level VI - evidence derived from a single descriptive or qualitative study. The main findings arranged in the objectives and conclusions are directly related to fetal heart disease and coping strategies for the health team (Table 1).

Given the above, it was possible to observe some factors related to the diagnosis of fetal congenital heart disease and the coping strategies for health teams.

## DISCUSSION

### Fetal heart disease and early diagnosis

CHDs are a developmental malformation of the heart structure that appears during the first weeks of pregnancy. They can range from simple conditions, which do not present symptoms, to complex situations, with more severe and potentially fatal symptoms, correcting themselves in the necessary time, configuring an early treatment.<sup>(14)</sup> It originates in the embryonic development of the cardiovascular system up to the 8th week of gestation and can be diagnosed in intrauterine life, by fetal echocardiography, from the 16th week of gestation. 1

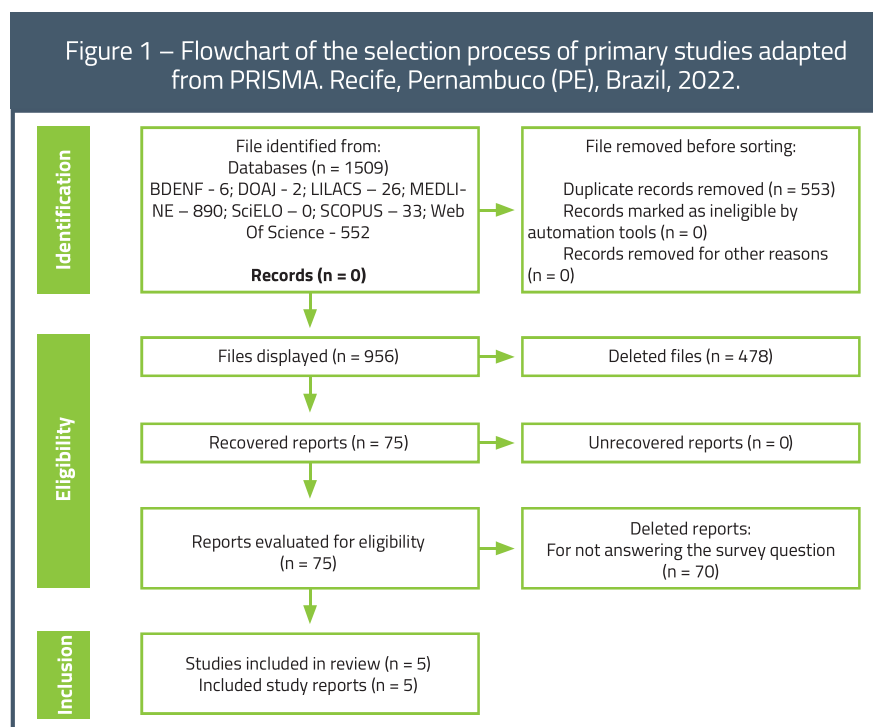
Currently, the real causes of congenital heart disease are not known, but some risk factors can be determined, including: Advanced maternal age (women over 35 years old); Cases in which the mother has diseases such as diabetes or lupus; 1st degree rela-

tive with congenital heart disease; Changes on morphological ultrasound, such as the presence of other suspected malformations or genetic syndromes; Consumption of alcohol, drugs and certain medications during pregnancy Maternal infections during pregnancy and multiple pregnancies and in vitro fertilization.<sup>(15)</sup>

Furthermore, Mozumdar et al.,<sup>(12)</sup> showed that factors associated with CHD included high anatomical complexity, maternal comorbidities, partners such as the

initial sonographer, and fewer fetal echocardiograms. As well, there are different types of congenital heart disease and each one has its own way of manifesting. The main signs and symptoms are: heart murmur, cyanosis (bluish discoloration of the skin and mucous membranes), fatigue on exertion, low weight gain, recurrent respiratory infections, arrhythmia (palpitations) and syncope (fainting). It can occur at any stage of life, from birth to adulthood.<sup>(16)</sup>

It is worth mentioning that prenatal



Source: Survey data, 2022.

Table 1 – Synthesis of the main findings about fetal heart disease and coping strategies for the health team. Recife, Pernambuco (PE), 2022.

N	Title/Database	Authors (Year)	Country	Level of Evidence	Objective	Results
1	Status of Multidisciplinary Collaboration in Neonatal Cardiac Care in the United States. / MEDLINE	Levy, Victor Y et al., (2021) <sup>(9)</sup>	USA	VI	Define the medical landscape of neonatal cardiac care and understand professionals' attitudes towards appropriate collaboration in this complex population within pediatric cardiac surgery programs.	Although outcomes for neonates with congenital heart disease have improved, it is evident that there is substantial variability between centers with regard to the multidisciplinary approach to care for this medically fragile patient population.

2	The pregnancy experience of Korean mothers with a prenatal fetal diagnosis of congenital heart disease. / MEDLINE	Im, YM et al., (2018) <sup>(10)</sup>	Korea	VI	To investigate the pregnancy experience of Korean mothers with prenatal fetal diagnosis of CHD	Early counseling with accurate CHD information, ongoing provision of clear explanations of prognosis, sufficient emotional support, and well-designed antenatal education programs are keys to an optimal outcome.
3	Screening for congenital heart defects: diversified strategies in current China. / Web Of Science	Liu, XW et al., (2019) <sup>(11)</sup>	China	VI	To review the use of diversified screening strategies in current China for the diagnosis of CHD.	Undoubtedly, it is more effective in improving the prognosis of patients if it is possible to discover and diagnose CHD in a timely manner, especially critical injuries, before birth, together with fetal interventions and perinatal management.
4	Diagnostic Accuracy of Fetal Echocardiography in Congenital Heart Disease / Web Of Science	Mozumdar, N et al., (2020) <sup>(12)</sup>	USA	II	To analyze the diagnostic accuracy of fetal echocardiography in congenital heart disease	Factors associated with CHD included high anatomical complexity, maternal comorbidities, partner as an initial sonographer, and fewer fetal echocardiograms.
5	Frequency of Congenital Heart Defects Detected on Fetal Echocardiography in High-Risk Mothers / Web Of Science	Iftikhar, Maryam; Hyder, Syed; Aziz, Saba (2016) <sup>(13)</sup>	Pakistan	IV	To determine the frequency of congenital heart defects on fetal echocardiography in high-risk mothers referred to Lahore Children's Hospital.	A 6% frequency of congenital heart disease was observed in high-risk mothers. With this high frequency of congenital heart defects, fetal echocardiography should be included as part of the second trimester anomaly examination in all high-risk mothers.

Source: Survey data, 2022.

care is essential for the diagnosis to occur, if there are suspected factors of fetal heart disorders. Since the diagnosis can be made even before the baby is born, with suspicion by morphological ultrasound and confirmation by fetal echocardiography of the maternity, with confirmation by other tests, especially electrocardiogram, Holter, chest X-ray, angiogram, cardiac catheterization and, especially, when using echocardiography (ultrasound of the heart), a non-invasive and relatively inexpensive method.<sup>(2,17)</sup>

Corroborating, Iftikhar, Hyder and Aziz<sup>(12)</sup> observed a 6% incidence of CHD in high-risk mothers. With this high frequency of CHD, it is suggested that fetal echocar-

diography should be included in the examination for second trimester abnormalities in all high-risk mothers.

Also, Liu et al.,<sup>(11)</sup> point out that it is highly effective to improve the patient's prognosis if CHD, especially critical injuries, can be detected and diagnosed early before birth, with fetal interventions and perinatal management.

**Coping strategies provided by health teams**

Treatment varies depending on the type and severity of heart disease. Some children progress to spontaneous recovery as they grow and develop, others require drug treatment, and a large proportion will requi-

re surgery or catheterization. Appropriate treatment should include multidisciplinary follow-up, which is composed of a psychologist, nutritionist, nurse, pediatric cardiologist, echocardiographer, cardiac surgeon, among others.<sup>(18)</sup>

Corroborating, Im et al.,<sup>(10)</sup> evidence in their study that early counseling with accurate information about CHD, continuous provision of clear prognostic explanations, adequate emotional support and well-designed prenatal education programs are the key to an ideal outcome.

Therefore, it was observed that although outcomes for newborns with CHD have improved, there are clearly significant differences between centers in the multidisci-

plinary approach to care for this medically fragile patient population.<sup>(9)</sup>

Through the search strategies, a small sample was raised. Although presenting a considerable result as a response to the crosses, few articles met the objective of the study. In addition, the included studies had limitations such as: different comparison systems and small sample size.

However, it was possible to highlight the possibilities of strategic coping to be provided by health teams to patients with CHD, as well as to pregnant women who received the diagnosis of fetal heart disease. However, it is still necessary to carry out

more studies containing a larger sample and enabling discussion about fetal heart disease and coping strategies.

This study can help to disseminate the importance of early diagnosis of congenital heart disease so that the follow-up and provision of adequate care can occur.

## CONCLUSION

This study highlighted the possibilities of strategic coping to be provided by health teams to patients with CHD, as well as to pregnant women who received a diagnosis of fetal heart disease through multidisciplinary

care. In addition to drug and surgical strategies, the healthcare team is responsible for early counseling with accurate CHD information, ongoing provision of clear prognostic explanations, adequate emotional support, and prenatal education programs.

However, there is a lack of studies that understand the true importance of this topic, essential in training, profession, and permanent education, even though this number has undergone a gradual increase in recent years. Therefore, this study showed an increase in the number of studies that carry out a survey of the strategies used in coping with CHD.

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