Original Article

Evaluation of Recent Updates Regarding the Diagnosis and Management of Congenital Heart in Children

Duaa Hamed Alahmadi[1*], Woroud Mohammad Alharthi[1], Albatol Taha Jifri[2], Mariyah Jaffer Alkhabbaz[2], Maram Khalid Alharbi[2], Hajar Ismail Alruwaili[3], Taha Jasim Almughais[4], Mohammed Suwaileh Alrehaili[5], Bayan Mohammed Hanif[6], Faris Nasser Alzaid[7], Dua'a Akram Rajh[7]

[1] Department of Medical Science, Faculty of Medicine, Batterjee Medical College, Jeddah, Saudi Arabia. [2] Department of Medical Science, Faculty of Medicine, Umm Al Qura University, Makkah, Saudi Arabia. [3] Department of Medical Science, Faculty of Medicine, jouf University, Aljouf, Saudi Arabia. [4]Department of Pediatric, Maternity and Children's Hospital, Dammam, Saudi Arabia. [5] Department of Pediatric, Ohud Hospital, Medina, Saudi Arabia. [6] Department of Medical Science, Faculty of Medicine, King Khalid University, Abha, Saudi Arabia. [7] Department of Medical Science, Faculty of Medicine, Almaarefa University, Riyadh, Saudi Arabia.

Abstract

Background: Congenital heart diseases are the most common cause of neonatal death worldwide, composing an array of various diseases and presentations. Early diagnosis and appropriate management are essential to the survival of children affected by this illness. **Objective:** Our aim was to identify the diagnostic approach and management of congenital heart disease as seen in the literature. **Methods:** PubMed database was used for articles selection, and the following keywords were used in the search: "congenital heart disease" and "evaluation". **Conclusion:** The condition, complications, and methods of preventing congenital heart disease should be carefully explained to parents, as it will increase their confidence as caretakers of their ill offspring. This helps reduce cardiovascular complications, hospitalizations, and mortality in CHD patients. Correct management of this illness is essential to the child's survival and multifactorial with regards to the type and severity of the condition.

Keywords: Congenital heart disease, management, evaluation, diagnosis, CHD

Introduction

Congenital heart disease (CHD) affects many newborns worldwide and continues to be a major burden on the fragile bodies of neonates [1]. Unfortunately, human cardiac lesions are unique in their irreproducibility in animal models, but fortunately for us is the advent of echocardiography and its ability to diagnose and secure knowledge on birth defects. Complacency in early diagnosis and management may have drastic consequences in the future. In this literature review of congenital heart disease, we discuss the evaluation of CHD through its prevalence in society and the world, the pathophysiology of its array of clinical manifestations, as well as focus on diagnostic approach and management. Our aim was to identify the diagnostic approach and management of congenital heart disease as seen in the literature.

Methodology

PubMed database was used for articles selection, and the following keywords were used in the MeSH (("Congenital Heart Disease"[Mesh]) AND ("Evaluation"[Mesh] OR "Management"[Mesh] OR "Diagnosis"[Mesh])). In regards to the inclusion criteria, the articles were selected based on the inclusion of one of the following topics; CHD evaluation, and CHD management and diagnosis. Exclusion criteria were all other articles, which did not have one of these topics as their primary endpoint, or repeated studies, and systematic reviews or meta-analysis.

Discussion

Congenital heart disease is a common cause of infantile death around the world, causing distress to mothers and caretakers. Knowledge of the many and common congenital heart defects

Address for correspondence: Duaa Hamed Alahmadi, Department of Medical Science, Faculty of Medicine, Batterjee Medical College, Jeddah, Saudi Arabia.
E-mail: d.h.s500 @ hotmail.com

How to cite this article: Hamed Alahmadi, D., Mohammad Alharthi, W., Taha Jifri, A., Jaffer Alkhabbaz, M., Khalid Alharbi, M., Ismail Alruwaili,H. and et al. Evaluation of Recent Updates Regarding the Diagnosis and Management of Congenital Heart in Children. Arch Pharma Pract 2019;10(3):29-32.

and their prevalence in the local community is necessary in order to prevent and manage this disease in a systematic and fruitful manner. Congenital anomalies may manifest in many forms and occasionally in the combination of deformities such as valvular defects and/or vascular abnormality. A heart murmur is a common clinical finding in CHD.

Epidemiology and Clinical Features

Ventricular septal defects are found on the outlet of the right ventricle, commonly it is a presentation of tetralogy of Fallot in around two-third of its occurrence, and in around 39% of the tetralogy of Fallot with pulmonary atresia [2, 3]. In Qassim, Saudi Arabia, the incidence of congenital heart defects was 5.4 per 1000 live births, with ventricular septal defects being the most common finding [4]. In Malaysia, it was reported that 6.7 of every 1000 live births had a form of congenital heart defect, developed countries in North America had 6.9 CHD cases per 1000 births, and a higher incidence was observed in European countries with 8.2 per 1000 births [5, 6]. In a study on the Chinese population, the high occurrence of congenital heart disease was associated with a maternal age above thirty-five years [7]. In Amorim et al. study, a third of CHD cases in newborns had isolated cardiac defect, a third had an associated birth anomaly, and around a quarter had cardiac abnormality as part of multiple anomaly syndrome. Having been diagnosed on autopsy or through Doppler echocardiography, the ventricular and atrial septal defects, along with patent ductus arteriosus were the most common anomalies found in both live and stillbirths [8]. Patent ductus arteriosus is related to decreased gestational age and along with other co-morbid conditions of prematurity. The ductus arteriosus is the vessel connecting the aorta and pulmonary trunk and lined with musculature, and while useful in balancing the intrauterine fetal circulation it becomes pathological post-delivery. Clinical finding depends on underlying illness, neonates with patent ductus arteriosus may present early with symptoms of poor feeding or increased oxygen demand, or they could remain asymptomatic and present later in childhood [9]. In aortic coarctation, there is a left-to-right shunting of blood flow in the presence of patent foramen ovale. These patients often end up in shock if the narrowing of the aorta occurs hastily, as no collateral circulation could have enough time to develop and counteract the impending congestive cardiac failure [10]. Eisenmenger patients are often associated with hemoptysis, the most common abnormality being a ventricular septal defect. Severe cardiovascular complications may be caused by Eisenmenger, from pulmonary embolisms, strokes, cerebral abscesses, and arrhythmias; eventually leading to sudden death and heart failure [11, 12]. The mindful state of patients with chronic diseases is an issue that should be investigated and managed early in the course of symptoms. Pregnant CHD patients may have spontaneous abortions, and when they successfully deliver these newborns may suffer from cyanotic CHD with cognitive impairment. Cognitive development difficulties are found in children with cyanotic congenital heart disease. In these scenarios, a neurodevelopmental strategy to reconstruct the mental, linguistic, and speech capabilities could be of great service to these children [13].

Diagnosis

There are many diagnostic modalities when it comes to approaching the cardiac defects in infants and children, yet echocardiography remains a cornerstone tool in diagnostic algorithms. Echocardiography is usually essential in diagnosing many congenital defects in live births including common ventricular and atrial septal defects and patent ductus arteriosus [8]. The feasibility and reproducibility of echocardiographic devices is a cornerstone in it being a preferred diagnostic modality in congenital heart diseases as its accessible and easily used in the clinical setting. However, the experience is essential for increased diagnostic accuracy and fastidious identification of anatomical abnormalities [14]. Endocrine disturbances are common in congenital heart disease systemic disturbances, notably subclinical hypothyroidism, which is associated with apparent cyanosis and younger age [15]. Hyperparathyroidism and low vitamin D levels were commonly found in children with metabolic bone disease and congenital heart anomalies, a population at high risk for skeletal fractures. [16]. Furosemide, a diuretic agent, was found to increase the possibility of fractures in children with congenital heart diseases, even when doses were non-continuous [17].

A meta-analysis of pulse oximetry usage in diagnosing CHD showed high diagnostic accuracy and specificity [18]. The importance of such a test is noted especially in a disease with no characteristic murmur like coarctation of the aorta in newborns, where a minority of cases could be detected by pulse oximetry. Aortic coarctation could be clinically diagnosed but is often misdiagnosed and patients may end up in shock. If newborns present with shock, and sepsis was ruled out, it is important to investigate aortic stenosis and coarctation as possible causes [10]. Fetal echocardiography may assist the physician in diagnosing coarctation, but does not replace clinical judgment [19]. Other modalities, such as transcatheter methods are leading in the diagnostic and therapeutic fields in patients with patent ductus arteriosus [20]. In CHD patients, testing for genetic abnormalities is usual, hence they may exhibit negative karyotyping but this should be revisited further by array genome testing. In a cohort sample of Saudi children with congenital heart disease, the novel genomic imbalances of duplication and deletions were identified by a test of microarray-based cytogenetics, these results of novel errors are encouraging their use within the first-line investigations in clinical diagnosis [21].

Management

Physical activity is a modality that was linked to arterial stiffness, with increased exercise associated with better health outcomes and reduced arterial stiffness in children, both of which are favorable outcomes [22]. These children with reduced physical activity have elevated base systolic blood pressure; hence, regular screening is needed to evaluate the need for treatment [23]. CHD not only causes issues at the

cellular level, but an abnormal increase in weight is observed in CHD children as they get older, increasing physical activity is a solution for both problems [24]. In infants with patent ductus arteriosus, a weight of fewer than 1000 grams at higher risk compared to their peers of normal weight. Prophylactic closure of a patent ductus arteriosus is no longer a recommendation, and the use of cyclooxygenase inhibitors would result in detrimental renal and mesenteric effects on the child. Oral ibuprofen in these children is associated with a lower risk of necrotizing enterocolitis [18]. Pulmonary hypertension is incurable morbidity, affecting around a tenth of CHD adult patients. Pulmonary hypertension in CHD patients requires adequate vasodilation therapy, phosphodiesterase 5 inhibitor (sildenafil) intake was associated with improved activity capacity and quality of life with microcirculatory benefits [25]. Elgendi et al. proposed a new clinical finding that could be used in diagnosing pulmonary arterial hypertension, this unique reduced first sinusoidal sound of entropy has significantly high specificity and sensitivity [26]. Vasodilatory therapies have been shown to reduce mortality in patients with Eisenmenger's syndrome [7].

Patients with one or more congenital heart diseases are at a higher risk of death and complications compared to their peers, in a follow-up study of postoperative cardiac correction surgery, the most frequent cause of mortality was adult re-operation or hospitalization for heart failure or ventricular tachyarrhythmia [27]. In children with depleting hemodynamic status, as should be seen with proper monitoring and screening, its advantageous, to their health, to implant cardioverter defibrillators, as post-resuscitation patients have demonstrated detrimental morbidity even after successful resuscitation [28]. Laparoscopic surgery is superior to open surgery, with regards to reduced blood transfusion requirements in all pediatric age groups, and especially beneficial in patients with mild CHD [29]. Post-operative cardiac catheterization is necessary for detecting and managing residual defects, this procedure could be performed safely in post-congenital heart surgery [30].

Conclusion

Congenital heart diseases continue to hold a psychological, economic, and physical burden on caretakers and patients, as the disease continues to corrode their health. The condition, its complications, and prevention should be carefully explained to parents as it would build their confidence as caretakers of their ill offspring. Emphasis on early diagnostic investigations and adherence to management plans should be sought. This helps in reducing cardiovascular complications, hospitalizations, and mortality in CHD patients. The early detection of these diseases is still on the physicians' attention so that they can chart their first management plan and provide optimal care from day one. These steps will provide a better prognosis and lifestyle. The new research points in CHD remains the new and upcoming breakthroughs in surgical techniques such as robotics surgeries as well as implementing the screening programs with genetic counseling.

References

1. Waggiallah HA. Thrombopoietin, Interleukin-11 And Soluble P-Selectin as Biomarkers in Children with Idiopathic Thrombocytopenic Purpura: Post-Steroid Therapy. International Journal of Pharmaceutical Research & Allied Sciences. 2018 Oct 1;7(4).
2. Mostefa-Kara M, Bonnet D, Belli E, Fadel E, Houyel L. Anatomy of the ventricular septal defect in outflow tract defects: similarities and differences. The Journal of thoracic and cardiovascular surgery. 2015 Mar 1;149(3):682-8.
3. Azzam ME, Mohamed AS, Shawky HA, Ahmed T, Nossier AM, Morcos MA, Ali MG. Early outcome of fallot repair with preservation of the pulmonary valve annulus versus transannular patch. Journal of Advanced Pharmacy Education & Research| Jan-Mar. 2018;8(1).
4. Al-Mesned A, Al Akhfash AA, Sayed M. Incidence of Severe Congenital Heart Disease at the Province of Al-Qassim, Saudi Arabia. Congenital heart disease. 2012 May;7(3):277-82.
5. Mat Bah MN, Sapian MH, Jamil MT, Abdullah N, Alias EY, Zahari N. The birth prevalence, severity, and temporal trends of congenital heart disease in the middle-income country: A population-based study. Congenital heart disease. 2018 Nov;13(6):1012-27.
6. van der Linde D, Konings EE, Slager MA, Witsenburg M, Helbing WA, Takkenberg JJ, Roos-Hesselink JW. Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. Journal of the American College of Cardiology. 2011 Nov 15;58(21):2241-7.
7. Sun PF, Ding GC, Zhang MY, He SN, Gao Y, Wang JH. Prevalence of congenital heart disease among infants from 2012 to 2014 in Langfang, China. Chinese medical journal. 2017 May 5;130(9):1069.
8. Amorim LF, Pires CA, Lana AM, Campos ÂS, Aguiar RA, Tibúrcio JD, Siqueira AL, Mota CC, Aguiar MJ. Presentation of congenital heart disease diagnosed at birth: analysis of 29,770 newborn infants. Jornal de pediatria. 2008;84(1):83-90.
9. J Vettukattil J. Pathophysiology of patent ductus arteriosus in the preterm infant. Current pediatric reviews. 2016 May 1;12(2):120-2.
10. Hoffman JI. The challenge in diagnosing coarctation of the aorta. Cardiovascular journal of Africa. 2018 Jul;29(4):252-5.
11. Arnott C, Strange G, Bullock A, Kirby AC, O'Donnell C, Radford DJ, Grigg LE, Celermajer DS. Pulmonary vasodilator therapy is associated with greater survival in Eisenmenger syndrome. Heart. 2018 May 1;104(9):732-7.
12. Daliento L, Somerville J, Presbitero P, Menti L, Brach-Prever S, Rizzoli G, Stone S. Eisenmenger syndrome. Factors relating to deterioration and death. European heart journal. 1998 Dec 1;19(12):1845-55.
13. Yilmaz İZ, Erdur B, Ozbek E, Mese T, Karaarslan U, Genel F. Neurodevelopmental evaluation of children with cyanotic congenital heart disease. Minerva pediatrica. 2018 Aug;70(4):365-70.
14. Miceli F. A review of the diagnostic accuracy of fetal cardiac anomalies. Australasian journal of ultrasound in medicine. 2015 Feb;18(1):3-9.
15. Bak P, Hjortshøj CS, Gæde P, Idorn L, Søndergaard L, Jensen AS. Subclinical hypothyroidism: A common finding in adult patients with cyanotic congenital heart disease. Congenital heart disease. 2018 Mar;13(2):263-70.
16. Cheng HH, Carmona F, McDavitt E, Wigmore D, Perez-Rossello JM, Gordon CM, Pigula FA, Laussen PC, Rajagopal SK. Fractures related to metabolic bone disease in children with congenital heart disease. Congenital heart disease. 2016 Jan;11(1):80-6.
17. Heo JH, Rascati KL, Lopez KN, Moffett BS. Increased Fracture Risk with Furosemide Use in Children with Congenital Heart Disease. The Journal of pediatrics. 2018 Aug 1;199:92-8.
18. Du C, Liu D, Liu G, Wang H. A meta-analysis about the screening role of pulse oximetry for congenital heart disease. BioMed research international. 2017;2017.
19. Lannering K, Bartos M, Mellander M. Late diagnosis of coarctation despite prenatal ultrasound and postnatal pulse oximetry. Pediatrics. 2015 Aug 1;136(2):e406-12.

20. Prescott S, Keim-Malpass J, Ikuta L, Zukowsky K. Patent ductus arteriosus in the preterm infant. Advances in neonatal care. 2017 Feb 1;17(1):10-8.
21. Al-Hassnan ZN, Albawardi W, Almutairi F, AlMass R, AlBakheet A, Mustafa OM, AlQuait L, Shinwari ZM, Wakil S, Salih MA, Al-Fayyadh M. Identification of novel genomic imbalances in Saudi patients with congenital heart disease. Molecular cytogenetics. 2018 Dec;11(1):9.
22. Boyes NG, Stickland MK, Fusnik S, Hogeweide E, Fries JT, Haykowsky MJ, Baril CL, Runalls S, Kakadekar A, Pharis S, Pockett C. Physical activity modulates arterial stiffness in children with congenital heart disease: A CHAMPS cohort study. Congenital heart disease. 2018 Jul;13(4):578-83.
23. Häcker AL, Reiner B, Oberhoffer R, Hager A, Ewert P, Müller J. Increased arterial stiffness in children with congenital heart disease. European journal of preventive cardiology. 2018 Jan;25(1):103-9.
24. Steele JM, Preminger TJ, Erenberg FG, Wang L, Dell K, Alsaied T, Zahka KG. Obesity trends in children, adolescents, and young adults with congenital heart disease. Congenital heart disease. 2019 Jan 30.
25. Clavé MM, Maeda NY, Thomaz AM, Bydlowski SP, Lopes AA. Phosphodiesterase type 5 inhibitors improve microvascular dysfunction markers in pulmonary arterial hypertension associated with congenital heart disease. Congenital heart disease. 2019 Mar;14(2):246-55.
26. Elgendi M, Bobhate P, Jain S, Guo L, Kumar S, Rutledge J, Coe Y, Zemp R, Schuurmans D, Adatia I. The unique heart sound signature of children with pulmonary artery hypertension. Pulmonary circulation. 2015 Dec;5(4):631-9.
27. Videbæk J, Laursen HB, Olsen M, Høfsten DE, Johnsen SP. Long-term nationwide follow-up study of simple congenital heart disease diagnosed in otherwise healthy children. Circulation. 2016 Feb 2;133(5):474-83.
28. Van Puyvelde T, Ameloot K, Roggen M, Troost E, Gewillig M, Budts W, Van De Bruaene A. Outcome after cardiopulmonary resuscitation in patients with congenital heart disease. European Heart Journal: Acute Cardiovascular Care. 2018 Aug;7(5):459-66.
29. Chu DI, Tan JM, Mattei P, Simpao AF, Costarino AT, Shukla AR, Rossano JW, Tasian GE. Outcomes of laparoscopic and open surgery in children with and without congenital heart disease. Journal of pediatric surgery. 2018 Oct 1;53(10):1980-8.
30. Nicholson GT, Kim DW, Vincent RN, Kogon BE, Miller BE, Petit CJ. Cardiac catheterization in the early post-operative period after congenital cardiac surgery. JACC: Cardiovascular Interventions. 2014 Dec 1;7(12):1437-43.