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Original Article

Prevalence of ophthalmological abnormalities in children and adolescents with CHD: systematic review and meta-analysis of observational studies

Manuel A. P. Vilela,[1] Graciele Sbruzzi,[2] Lucia C. Pellanda[3,4]

[1]*Ophthalmology Department, Universidade Federal de Pelotas, Pelotas;* [2]*School of Physical Education, Universidade Federal do Rio Grande do Sul;* [3]*Post Graduation Program in Health Sciences: Cardiology, Instituto de Cardiologia/Fundação Universitária de Cardiologia;* [4]*Universidade Federal de Ciências da Saúde de Porto Alegre, Porto Alegre, RS, Brazil*

Abstract *Background:* CHDs form a complex and heterogeneous group of clinical entities, with high morbidity and mortality. With the advancement of surgical and invasive techniques and clinical treatment, the survival of these patients has increased significantly, and there are reports of a high prevalence of ocular abnormalities in this group. The objective of this study was to estimate the prevalence of ocular findings in children and adolescents diagnosed with CHD. *Methods:* A systematic search was conducted in the following databases: MEDLINE (via PubMed), EMBASE, and Cochrane CENTRAL, in addition to a manual search on studies published on the patient, from inception until August, 2014. Observational studies assessing the prevalence of ocular abnormalities in children and adolescents with CHDs were included. *Results:* Of the 2413 articles identified, eight were included, comprising a total of 1061 patients. Among them, the lowest and highest prevalences observed were 6.3 and 65%, respectively. The weighted average prevalence of ocular abnormalities was 32.5% (CI95% 19.3–49.3). Strabismus, cataracts, and retinopathy were the most frequently observed alterations. *Conclusion:* The prevalence of ocular abnormalities in children and adolescents with CHDs was 32.5%, demonstrating that ocular consequences are not uncommon in this population and may have relevant clinical impact. These results reinforce the need for ophthalmological evaluation of patients with CHDs.

Keywords: Eye diseases; CHDs; review; meta-analysis

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HEART DISEASES ARE AMONG THE MOST COMMON congenital diseases, occurring in 1% of live births, with frequent co-existence of ocular sequelae.[1] These consequences include retinal arteriolar narrowing (focal and diffuse), increased vascular tortuosity, retinal capillary disease – found as oedema or ischaemic and haemorrhagic infarction – thrombotic or embolic occlusion of the retinal artery, choroid and optic disc, and proliferative retinopathy with secondary retinal detachment. Other signs of ocular involvement include cataracts, strabismus, glaucoma, eyelid abnormalities, and amblyopia, with or without associated genetic syndromes.[2–7] The prevalence of ocular signs in patients with CHDs ranges from 6.3[8] to 65%[9] in literature reports. Such a large variation may be due to sample characteristics, heterogeneity of heart diseases, and techniques of ophthalmic evaluation. In addition, most of the published data include case reports or case series with specific heart lesions, with scarce information from population studies. Our hypothesis is that true prevalence may be closer to the higher values, as the largest and methodologically sound studies report

Correspondence to: M. A. P. Vilela, MD, PhD, Department of Specialized Medicine – Ophthalmology, Federal University of Pelotas, Avenida Duque de Caxias, 250, Fragata, Pelotas, RS 96001970, Brazil. Tel: +55 533 309 2400; Fax: +55 5133953602; E-mail: mapvilela@gmail.com

high prevalences, and survival has been increasing progressively in more complex patients.

Thus, the present study aimed to estimate the prevalence of ocular abnormalities in patients from birth to adulthood with CHD, through a systematic review with meta-analysis of observational studies.

Methods

Sources and search strategy

The investigators, who received formal training in systematic review, performed all searches. A systematic search was performed in the following electronic databases: MEDLINE (accessed via PubMed), Cochrane Central Register of Controlled Trials (Cochrane CENTRAL), and EMBASE, from inception until August, 2014. In addition, references of studies published on the patient were manually searched and authors were contacted when necessary. The search strategy used in PubMed is shown in Table 1. There was no language restriction. One non-English paper was translated by an experienced professional.[10] A manual search was performed, but no unpublished study or conference abstract fulfilled the inclusion criteria. Thus, there was no need to contact authors for further information or to handle unpublished abstracts.

Eligibility criteria

This review included observational studies – cohort, cross-sectional, or case–control studies – baseline results of randomised or non-randomised clinical trials, or observational follow-up after clinical trials assessing the prevalence of ocular findings in children and adolescents aged between 6 and 18 years with CHDs. The primary outcome was the presence of ocular symptoms, including all findings detected in the eyeball and annexes.

Table 1. Search strategy used for the PubMed database.

#1	"Heart Septal Defects, Ventricular"[Mesh] OR "Ventricular Septal Defect" OR "Ventricular Septal Defects" OR "Defect, Ventricular Septal" OR "Septal Defect, Ventricular" OR "Septal Defects, Ventricular" OR "Intraventricular Septal Defects" OR "Intraventricular Septal Defect" OR "Heart Septal Defects, Atrial"[Mesh] OR "Atrial Septal Defects" OR "Defect, Atrial Septal" OR "Septal Defect, Atrial" OR "Septal Defects, Atrial" OR "Atrial Septal Defect" OR "Persistent Ostium Primum" OR "Ostium Secundum Atrial Septal Defect" OR "Aortic Coarctation"[Mesh] OR "Aortic Coarctations" OR "Coarctation, Aortic" OR "Fontan Procedure"[Mesh] OR "Fontan Operation" OR "Hemi-Fontan Procedure" OR "Hemi Fontan Procedure" OR "Bidirectional Glenn Shunt" OR "Bidirectional Glenn Shunts" OR "Bidirectional Glenn Procedure" OR "Bidirectional Glenn Procedures" OR "Bidirectional Cavopulmonary Shunt" OR "Bidirectional Cavopulmonary Shunts" OR "Tricuspid Atresia"[Mesh] OR "Tricuspid Atresias" OR "Absent Right Atrioventricular Connection" OR "Tricuspid Valve Atresia" OR "Tricuspid Valve Atresias" OR "Right ventricle hypoplasia" OR "Right ventricular hypoplasia" OR "Isolated right ventricular hypoplasia" OR "Hypoplasia of the right ventricle" OR "Isolated hypoplasia of the right ventricle" OR "Transposition of Great Vessels"[Mesh] OR "Great Vessels Transposition" OR "Transposition of Great Arteries" OR "Great Arteries transposition" OR "Tetralogy of Fallot"[Mesh] OR "Fallot's Tetralogy" OR "Fallot Tetralogy" OR "Fallots Tetralogy" OR "heart defects, congenital"[mesh] OR "congenital heart defect" OR "heart abnormalities" OR "heart abnormality" OR "congenital heart"
#2	"Visual Acuity"[Mesh] OR "Acuities, Visual" OR "Acuity, Visual" OR "Visual Acuities" OR "acuities" OR "Amaurosis Fugax"[Mesh] OR "Monocular Blindness, Transient" OR "Blindness, Transient Monocular" OR "Transient Monocular Blindness" OR "Blindness, Monocular, Transient" OR "Blindness"[Mesh] OR "Blindness, Acquired" OR "Acquired Blindness" OR "Blindness, Monocular" OR "Monocular Blindness" OR "Blindness, Hysterical" OR "Hysterical Blindness" OR "Blindness, Transient" OR "Transient Blindness" OR "Blindness, Legal" OR "Legal Blindness" OR "Amaurosis" OR "Amauroses" OR "Blindness, Complete" OR "Complete Blindness" OR "Visual Perception"[Mesh] OR "Perception, Visual" OR "Perceptions, Visual" OR "Visual Perceptions" OR "Visual function" OR "Vision, Ocular"[Mesh] OR "Ocular Vision" OR "Vision" OR "Light Signal Transduction, Visual" OR "Visual Phototransduction" OR "Phototransduction, Visual" OR "Visual Transduction" OR "Transduction, Visual" OR "Visual Light Signal Transduction" OR "Retina"[Mesh] OR "retinas" OR "Retinopathy" OR "Retinal Diseases"[Mesh] OR "Disease, Retinal" OR "Diseases, Retinal" OR "Retinal Disease" OR "Retinal vascular disease" OR "Retinal Vessels"[Mesh] OR "Retinal Vessel" OR "Vessel, Retinal" OR "Vessels, Retinal" OR "Retinal Blood Vessels" OR "Blood Vessel, Retinal" OR "Blood Vessels, Retinal" OR "Retinal Blood Vessel" OR "Vessel, Retinal Blood" OR "Vessels, Retinal Blood" OR "Pecten Oculi" OR "Vascular tortuosity" OR "Retinal vascular occlusion" OR "Stasis retinopathy" OR "Retinopathy of Prematurity"[Mesh] OR "Prematurity Retinopathies" OR "Prematurity Retinopathy" OR "Retrolental Fibroplasia" OR "Fibroplasia, Retrolental" OR "Fibroplasias, Retrolental" OR "Retrolental Fibroplasias" OR
#2	"Refractive Errors"[Mesh] OR "Error, Refractive" OR "Errors, Refractive" OR "Refractive Error" OR "Refractive Disorders" OR "Disorder, Refractive" OR "Disorders, Refractive" OR "Refractive Disorder" OR "Ametropia" OR "Ametropias" OR "Eye Injuries"[Mesh] OR "Eye Injury" OR "Injury, Eye" OR "Injuries, Eye" OR "eye lesion" OR "Amblyopia" OR "Amblyopia"[Mesh] OR "Vision, Binocular"[Mesh] OR "binocular vision" OR "Glaucoma"[Mesh] OR "Glaucomas" OR "Strabismus"[Mesh] OR "Squint" OR "Phorias" OR "Phoria" OR "Strabismus, Noncomitant" OR "Noncomitant Strabismus" OR "Mechanical Strabismus" OR "Strabismus, Mechanical" OR "Strabismus, Comitant" OR "Comitant Strabismus" OR "Convergent Comitant Strabismus" OR "Comitant Strabismus, Convergent" OR "Strabismus, Convergent Comitant" OR "Hypertropia" OR "Hypertropias" OR "Cataract"[Mesh] OR "Cataracts" OR "Lens Opacities" OR "Lens Opacity" OR "Opacities, Lens" OR "Opacity, Lens" OR "Cataract, Membranous" OR "Cataracts, Membranous" OR "Membranous Cataract" OR "Membranous Cataracts" OR "Pseudoaphakia" OR "Pseudoaphakias" OR "Optic nerve" OR "Optic nerve injuries" OR "Optic Disk"
#3	#1 AND #2

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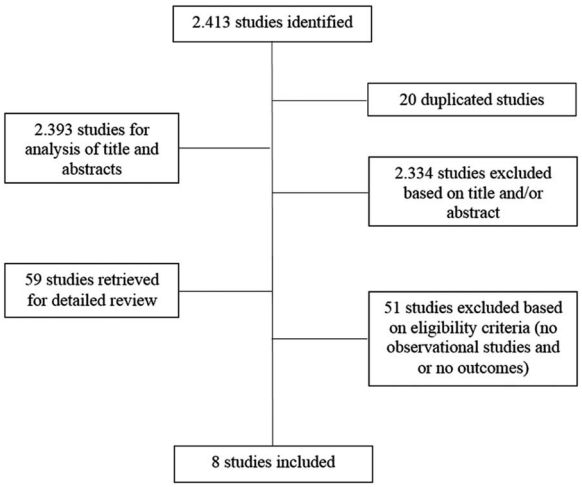

Figure 1.
Flow chart of the studies included in this review.

Selection of studies and data extraction

The analysis of titles and abstracts of all publications identified in the search strategy, as well as the extraction of the data regarding the methodological characteristics of the studies, patients, and results, were carried out in duplicate by two independent reviewers – L.C.P and M.A.P.V. – through standardised forms. Papers whose abstracts did not contain sufficient information regarding the inclusion and exclusion criteria were selected for full evaluation. In the second phase, the same reviewers independently assessed the complete articles and selected them in accordance with the eligibility criteria. The extracted variables included all signs of structural or functional modifications involving the eyeball and its annexes. Discrepancies between the results from the two reviewers were resolved by consensus and, in cases of persistent differences, by a third assessment (G.S.).

Assessment of risk of bias

For the assessment of risk of bias, two independent and blinded reviewers – L.C.P and M.A.P.V. – assessed the methodological quality of the studies, based on the criteria of Downs and Black,[11] which considers the following 5 items: available information – objectives, outcome, sample description, description of loss, variability of results, and the actual probability of the findings – external validity – representativeness of the sample, sampling procedures, site representativeness, and team involved in the intervention – bias – kind of blinding, prior planning of analysis, duration of follow-up, adequacy of analysis, and accuracy of the tests – confusion – origin of the population, selection period, randomisation, allocation, adjusted analysis and analysis for intention to treat, and critical appraise of losses in the discussion section – and power of the study.

Disagreements between the reviewers were resolved by consensus and, in cases of persistent disagreement, by a third reviewer (G.S.).

Data analysis

The quantitative analysis of the included studies was performed by meta-analysis of single arm, with estimation of an average prevalence weighted by sample size. A random effect model was used. The meta-analysis was performed in the *Comprehensive Meta Analysis* software. Heterogeneity between studies was explored using the following strategy: we repeated the meta-analyses, removing one paper at a time to check whether an individual study explained heterogeneity.

Results

Description of studies

Of the 2413 studies identified in the search, eight matched the eligibility criteria, yielding a total of 1061 patients with CHDs; one of the studies also had a control group composed of 76 healthy individuals.[8] Figure 1 shows the flowchart of the studies included in this review. The age of the individuals ranged from 6 days to 39 years. All the included articles were cross-sectional studies; four of the studies[1,8,12,13] examined the prevalence of ocular findings in patients with all forms of CHD; three[10,14,15] only in cases with coarctation of the aorta; and one study[9] only in patients with cyanotic heart disease. The characteristics of included studies are shown in Table 2.

Assessment of risk of bias

To assess the methodological quality of the studies, the criteria for experimental studies were excluded from the score, as no study with this design was selected. Thus, the maximum score for the 17 items was 12 points (minimum 7 and maximum 12, mean 9.3 points). The average score in the included studies was 7.25/12 (SD $\pm$ 1.83) possible points (75% with 6 points, 12.5% with 7, and 12.5% with 8 points) (Table 3).

Prevalence of ocular findings

The lowest prevalence found in the included studies was of 6.3%[8] and the highest was 65%.[9] The average sample-size weighted prevalence was 32.5% (CI95% 19.3–49.3%) (Table 2 and Fig 2).

Studies with all forms of CHD. The prevalence of ocular manifestations in patients with CHDs not associated with syndromes was 6.6–32.6%.

Alfano[12] evaluated 500 sequential cases of children with CHD not associated with rubella, aged between 6 days and 15 years. Excluding patients with genetic

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Table 2. Characteristics of the studies included in this review.

Reference	Type of study	Type of CHD	Age	Sample size (n)	Ocular abnormalities	Prevalence (%)
Alfano[12]	Cross-sectional	Non-specified CHD	6 days–15 years	478	Cataract 0.5%, glaucoma 0.4%, strabismus 1.6%, microphthalmia 0.8%, ptosis 0.4%, obstruction of lacrimal duct 0.4%, coloboma 0.8%	10.4
Gardiner and Joseph[13]	Cross-sectional	Patients with cyanotic, acyanotic lesions, with left-to-right shunts, and obstructive lesions.	6–15 years	85	Strabismus 14%, coloboma 1.1, amblyopia 22.4, retinal vascular dilatation 6.3	17.6
Fusco et al[8]	Cross-sectional	20 CCHD 35 right-to-left shunts 21 obstructive heard disease	2–39 years	76	Retinal vascular dilatation 6.3, optic nerve swelling 1.3	6.3
Mansour et al[1]	Cross-sectional	37.5% with volumetric overload or right-to-left shunts, 36.3% cyanotic and 26.3% obstructive	2.9 ± 4.2	240	Cataract 0.75, strabismus 1.48, ptosis 0.78, optic nerve hypoplasia 9.6, increased retinal vascular tortuosity 15.5, retinal haemorrhages 4.4	32.6
Eisalo et al[14]	Cross-sectional	ACo	17–46 years	25	Retinal vascular narrowing 20, increased retinal vascular tortuosity 24, capillary dilatation 40	36
Johns et al[15]	Cross-sectional	ACo	9 days–20 years	20	Retinal vascular narrowing 10, increased tortuosity 20, retinal haemorrhages 5	45
Raczyńska et al[10]	Cross-sectional	ACo	Mean = 16 years	54	Anomalous arteriovenous connections 19, increased dorsal reflex of the vessels 21, increased tortuosity 35	29.6
Petersen and Rosenthal[9]	Cross-sectional	CCHD	2–26 years	83	Retinal vascular dilatation 20.5, optical nerve oedema 14	35

Table 3. Risk of bias of included studies.

Reference	Study quality	External validity	Bias	Confusion	Sample power	Downs and Black mean score
Alfano[12]	Adequate	Inadequate	Adequate	Unclear	Adequate	7
Gardiner and Joseph[13]	Adequate	Adequate	Adequate	Inadequate	Inadequate	8
Eisalo et al[14]	Adequate	Inadequate	Adequate	Inadequate	Inadequate	5
Petersen and Rosenthal[9]	Adequate	Adequate	Adequate	Inadequate	Inadequate	8
Fusco et al[8]	Inadequate	Inadequate	Adequate	Inadequate	Inadequate	5
Johns et al[15]	Adequate	Inadequate	Adequate	Inadequate	Inadequate	6
Raczyńska et al[10]	Adequate	Adequate	Adequate	Inadequate	Inadequate	9
Mansour et al[1]	Adequate	Adequate	Adequate	Inadequate	Adequate	10
						7.25

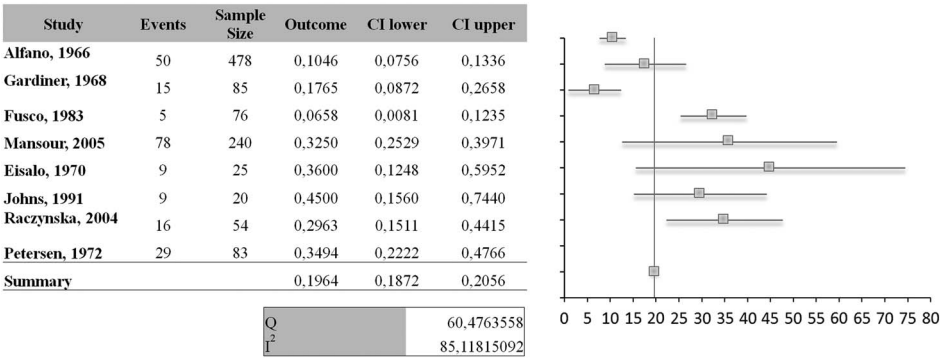

Figure 2.
Prevalence of ocular findings in children and adolescents with CHD.

syndromes, with Down or Hurler syndrome, ocular manifestations were detected in 52 children (10.4%). Scleral cyanosis was observed in 32% of them. Although the prevalence of cataract (0.50%) and glaucoma (0.40%) was lower than rubella, it was much higher than that in the general population. Strabismus was present in ~1.6% of the patients, microphthalmia in 0.8%, ptosis in 0.4%, coloboma in 0.8%, and obstruction of the lacrimal ducts in 0.4% of the cases. The type of heart disease presented by the patients or the routine exams used was not described in this study.

Gardiner and Joseph[13] analysed 85 children with CHDs (age with a range between 6 and 15 years) seen within a period of 12 months. The global prevalence of non-refractive ocular findings was 17.6% (number of cases: strabismus, 12; cataract, 2; coloboma of choroid, 1). Ocular alterations were observed in 19 of 22 cyanotic children (86%), 18 of 28 obstructive patients (64%), and 17 of 35 children with shunts (49%). The frequencies of amblyopia and of ametropia were 23.5 and 36%, respectively.

Fusco et al[8] examined 76 patients, aged between 2 and 39 years, with CHD. Fundoscopic alterations were found in 6.3% of the patients, precisely in three cases with cyanosis and in two of the patients with aortic stenosis. Symptoms observed in these five patients included vascular dilatation with bluish-coloured blood vessels and, in only one patient with Fallot (1.3%), optical disc oedema. After surgery to correct heart defects, functional changes were seen in 18.4% of campimetry and 37.7% of chromatic tests.

Mansour et al[1] studied 240 cases (mean age 2.9 years; SD = 4.2 years). The most common cardiac abnormalities were atrial or ventricular septal defects (n = 62), tetralogy of Fallot (n = 39), pulmonary stenosis (n = 25), and transposition of the great arteries (n = 24). Other forms found were as follows: patent arterial duct (n = 13), double right ventricle (n = 11), aortic stenosis (n = 10), pulmonary atresia (n = 10), coarctation of the aorta (n = 9), single ventricle (n = 6), and atrioventricular canal defect (n = 6). Exclusion of patients with genetic syndromes reduced the sample to 135 cases (56.25%), with a prevalence of ocular findings of 32.6%. In this specific group, the ocular findings were as follows: ptosis (0.75%), congenital cataracts (0.75%), strabismus (1.48%), retinal haemorrhages (4.4%), optic nerve hypoplasia (9.6%), and retinal arterial and venous tortuosity (15.5%). The prevalence of ptosis, strabismus, and cataract was significantly higher in cases associated with syndromes, but the distribution of results depending on the specific type of anomaly among patients with CHDs was not described.

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Patients with aortic coarctation. In studies evaluating patients with coarctation of the aorta, the prevalence of ocular abnormalities ranged from 29.6 to 45%.

In a cross-sectional study, Eisalo et al[14] evaluated by angiofluoresceinography 25 cases with coarctation of the aorta. The patients aged between 17 and 46 years, and 60% of them were male. Mild or severe abnormalities were recognised in 36% of the patients, and included arterial narrowing (20%), arterial and venous tortuosity (24%), and increased capillary visibility (40%).

In addition, in a cross-sectional study, Johns et al[15] examined 20 cases with coarctation of the aorta. The patients aged between 9 days and 20 years, and 70% of them were male. The prevalence of ocular abnormalities was 45%, with 5% of retinal haemorrhages, 10% strabismus, 15% retinal venous and arterial tortuosity, and 20% retinal arterial tortuosity only.

Raczyńska et al[10] investigated a series of 54 cases (mean age of 16 years) with coarctation of the aorta and arterial hypertension. Ocular abnormalities were observed in 29.6% of the patients, including arteriovenous crossings in 19%, increased dorsal reflex in 21%, increased venous tortuosity in 26%, vascular narrowing in 28%, and anomalous arterial tortuosity in 35% of the cases.

Patients with cyanotic CHD. Petersen and Rosenthal[9] analysed 83 patients aged between 2 and 26 years with congenital cyanotic heart disease in a cross-sectional study. Moderate-to-severe fundoscopic abnormalities were found in 35% of the patients, with 14.45% of them presenting venular dilatation with disc oedema, and 20.5% presenting isolated retinal venular dilation.

Discussion

In this systematic review with meta-analysis, we found that 32.5% (CI95% 6.3–65%) of patients with CHDs not associated with specific syndromes have ocular abnormalities. These alterations were more prevalent and more severe among patients with cyanotic CHDs.

It is possible that cataract, strabismus, colobomas, retinopathy, and amblyopia have higher prevalence among patients with CHDs compared with the normal population.[2,16–20] Most of the abnormalities are seen in the retina, including venular dilation, arterial and venous tortuosity, arteriolar narrowing, increased arterial dorsal reflex, anomalous arteriovenous connections, micro-haemorrhage, and optic nerve swelling.[2,4,7,21–27]

The population prevalence of congenital cataract is 1.2–6 cases/10,000 individuals (0.012–0.06%).[18] In the studies included in this review, this prevalence ranged from 0.5 to 2.3%. Clearly, this form of cataract was more incident, but the reason is not known. Strabismus was reported in 1.5–14% of patients with CHDs, whereas in the general population its prevalence is 2.1–3.3%.[16,20] This wide range may suggest a possible trend, but there is no definitive evidence for a relationship. The overall prevalence of ocular coloboma is 0.0024%.[12,13,17] Alfano[12] and Gardiner and Joseph[13] detected this abnormality in 0.8–1.1% of the patients with CHDs, but this relationship was not described in the other studies selected in this systematic review. Owing to the lack of population data or to the small sample size, a relationship between CHDs and other conditions described, such as congenital ptosis, microphthalmia, and obstruction of lacrimal duct, was not possible to investigate in the studies.

Retinal changes related to CHDs include increased dorsal arterial reflex, pathologic arteriovenous anastomosis, vascular narrowing or dilatation, increased arterial and/or venous tortuosity, and retinal haemorrhages. Vascular tortuosity was the most commonly described of these findings, ranging from 15.5 to 35% of the patients. An important correlation was observed between presence of coarctation of the aorta and tortuosity (average 26.3%). It is known that this abnormal change in vascular path increases with age, with a relationship with the diameter of the vessel and its transmural pressure.[2,28] The pressure elevation initially modifies the vessel's diameter, but if critical levels are reached the route of the vessel may be affected. It is probable that the retinal findings are directly related to factors such as hypoxia, blood hyperviscosity, and high blood pressure.[21,22,29] Individually or as a group, these factors can induce these changes, and may even affect the capillary network, with transudation and/or occlusion. Many of these signs are biased by the evaluator's subjectivity and have low diagnostic sensitivity (3–21%). In case of systemic arterial hypertension, however, the findings have high specificity (88–98%). Therefore, hypertensive retinopathy may not be common in patients with chronic systemic arterial hypertension, but is rare in normotensive individuals. The presence of hypertensive retinopathy increases significantly the risk for left ventricular hypertrophy (odds ratio = 2.2), which is typical in patients with coarctation. Furthermore, cerebrovascular accident is the only cardiovascular event strongly linked to hypertensive retinopathy.[24]

The information obtained from studies on neurological damage associated with CHDs shows evidence of a delay in the volumetric and functional development, myelination, and formation of convolutions and glial bands in the central nervous system in 20% of the patients.[29] As the retina has the same embryological origin, and therefore shares many common

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aspects with the central nervous system, it is possible that in addition to anatomical changes, functional ocular abnormalities are a consequence of CHDs.

Some limitations of this systematic review merit discussion. The small number of studies limited subgroup or sensitivity analyses. As in all systematic reviews, one major concern is the publication bias. As there is no registry of observational studies that could serve as a source of unpublished work in the same format as the clinical trials or systematic reviews registries, there is always a possibility of remaining bias. On the other hand, some methodological strengths could have served as strategies to decrease the possibility of this bias. First, we selected key words and conducted the strategy search to get most sensitive citations selection. Second, the overall references screening and the inclusion/exclusion paper criteria were carried out with no language restrictions, as there is a trend of publishing positive results in English and this may increase publication bias (or English bias). Third, we used all the efforts to locate unpublished studies, performing manual searches in key periodicals and references.

Another possible limitation is the high heterogeneity between studies. Several methodological reasons may help in explaining this heterogeneity. They are related mainly to the type of examination (full or only partial) and resources used for the evaluation. The description of these results depends on the different equipment or techniques used: in the case of fundoscopy, for example, studies with and without mydriasis, using direct or indirect binocular ophthalmoscope, or even through electroretinogram with different fields. Combining all the studies included in this review, a total of 1061 patients were included; however, most samples were small and were collected in very different times relating to surgical or percutaneous correction. This may result in an overestimation of ocular abnormalities in the earliest studies, as associated syndromes may not have been well diagnosed or excluded. The high heterogeneity of results reported in these studies may be related to differences in evaluation methods, as already mentioned, but also to the heterogeneity of CHDs themselves, which form a group of very different entities, both in pathophysiology and in severity.

In summary, different cardiovascular conditions described in the literature are associated with ocular effects. Strong evidence shows that hypertension modifies the state of the retinal vessels from narrowing to occlusion and haemorrhages or severe oedema.[2,24,25] Heart failure, ischaemic heart disease, cardiac valve disorders, aortic arch disease, and carotid stenosis are conditions where different retinal signals are described.[21–26] Another possible link is the presence of genetic abnormalities that are associated with cardiac and ophthalmic alterations, even if not resulting in specific syndromes. As CHD is associated with many of these mechanisms, including modifications in perfusion pressure, secondary hypoxia, and thromboembolic events, the presence of ocular signs is to be expected and maybe actively investigated.

In this way, the present systematic review with meta-analysis allows the conclusion that at least one-third of patients with CHDs, not linked to genetic syndromes, present ocular abnormalities. These findings were more prevalent, although not pathognomonic, in patients with cyanotic CHDs. Cataract, strabismus, and retinopathy were the most frequently observed abnormalities, with a prevalence higher than that in the normal population. The identification of these ocular manifestations has diagnostic and prognostic significance, allowing the implementation of more effective strategies to prevent morbidities in adulthood.

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Conflicts of Interest

None.

Ethical Standards

The authors assert that all work reported complies with the ethical standards of the Helsinki convention, and Ethics Board of the Institute of Cardiology, Porto Alegre, Brazil.

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