

DIADNOSIS, MANAGEMENT AND PROGNOSIS OF PATIENTS WITH CRITICAL COARCTATION OF THE AORTA

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ABSTRACT

The aim of this study is to analyze the clinical presentation, diagnosis, operative results and prognosis of the patients with critical coarctation of the aorta (CoA).

METHODS AND MATERIALS: A group of 43 newborns and infants up to 3 months of age with critical CoA out of 272 patients with critical congenital heart diseases (CCHD) from the region of North-East Bulgaria were retrospectively followed for a period of 25 years (1987-2011). Methods of investigation included: clinical examination, electrocardiography, conventional radiography, echocardiography and cardiac catheterization. Corrective procedures were done in 31 of the children. Time of postoperative follow-up was an average of 6 years. Operating results are evaluated on a scale developed by us.

RESULTS: The CoA is the second most frequent CCHD after the transposition of the great arteries (TGA) - 16%, most often in conjunction with other cardiac abnormalities. The clinical presentation and the time of diagnosis of the patients were significantly delayed ($p < 0,001$). One third of newborns with CCHD, discharged from hospital without being diagnosed with cardiac malformation, turned out to be with CoA. Only 9.5% of our patients were diagnosed in neonatology department, versus 32.7% in other critical cardiopathies ($p < 0,001$) and in one fourth of the children the diagnosis was incorrect or incomplete. Corrective surgical procedures were performed in 31 (72.1%) of the children – 1.6 intervention on each patient average. In 64.5% one-stage correction was performed with spontaneously recovery of the concomitant heart malformation. The preferable surgical technique was Amato procedure – resection and plastic "end-to-end". Early postoperative mortality was 9.7%. Reoperations were performed in 6 children - 19.4 percent. The late operating results are often good or very good in 65.4% of patients.

CONCLUSION: CoA is a common CCHD and the time of the clinical manifestation and the diagnosis are significantly delayed. In spite of the common concomitant heart anomalies in most of the patients it is possible a one-stage corrective procedure to be performed with good postoperative result.

Key words: *critical coarctation of the aorta, critical congenital heart disease, clinical manifestation, diagnosis, surgical results, prognosis*

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Received: December 18, 2013

Accepted: January 10, 2013

Coarctation of the aorta (CoA) is a relatively often cardiac malformation – 5-8% of congenital heart diseases (CHD). It is a segmental narrowing, including thickening of the media and accumulation of neonatal tissue, most often in the isthmal part of the aorta. The neonatal type is usually associated with some other cardiac defects as patent ductus arteriosus (PDA), ventricular septal defects (VSD), aortic stenosis (AS) and some rare malformations of the

mitral valve. The critical CoA in the neonatal period and early infancy is presented with cardiogenic shock, metabolic acidosis and organ dysfunction or congestive heart failure (5). It is connected with the passability of the arterial duct, severity of obstruction, accompanying malformations, working compensatory mechanisms and decreased capability of the neonatal myocardial tissue (7). There is an urgent surgical or interventional therapy without the patients died.

The aim of this study is to analyze the clinical presentation, diagnosis, operative results and prognosis of the patients with critical CoA.

METHODS AND MATERIALS

Table 1. Concomitant cardiac malformations in patient with critical CoA

Concomitant CHD	VSD	AS/BAoV	MS	Hypoplastic Ao arch	Other	All
Count	16	13	4	6	4	43

A group of 43 newborns and infants up to 3 months of age with critical CoA out of 272 patients with critical congenital heart diseases (CCHD) from the region of North-East Bulgaria were retrospectively followed for a period of 25 years (1987-2011). Methods of investigation included: clinical examination, electrocardiography, conventional radiography, echocardiography and cardiac catheterization. Corrective procedures was done in 31 of the children. The surgical results are described as very good, good, satisfactory and unsatisfactory based on the clinical condition of the patients, hemodynamic indices, rhythm and conduction disorders, extracardiac postoperative complications and the need of reoperation.

RESULTS AND DISCUSSION

In our study the CoA is the second most frequent CCHD after the transposition of the great arteries (TGA) - 16% (Figure 1).

The rationing by sex is nearly the same – 21 boys and 22 girls. Forty-three concomitant cardiac malformations were established: VSD – 16, AS/bicuspid valve – 13, hypoplastic aortic arch – 6, mitral valve stenosis – 4 and some other rare like Shon’s syndrome, aortopulmonary fenestration, anomalous

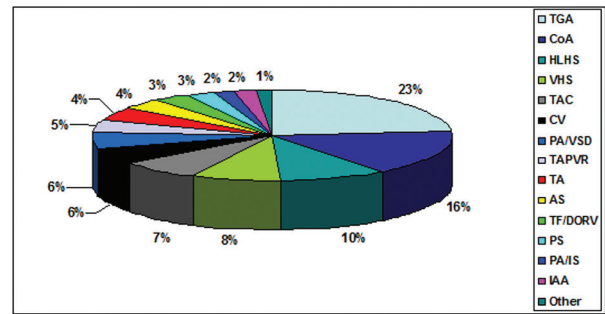


Fig. 1. Nosological structure of CCHD.

right pulmonary artery from the aorta and hypoplasia of the ascending aorta. In some of the cases the malformations are hemodynamically insignificant and practically this means isolated CoA (Table 1).

Extracardiac malformations and syndromes were found in 11 (25.6%) of the children, but they were not more frequent than those in the whole CCHD group – 30%.

The time of initial clinical presentation of CoAo is at average 7.1 days (95% CI: 4,9-9,4) versus 3.4 days (95% CI: 2,7-4,2) in the whole CCHD group and the age of diagnosis establishment of CoA is 15.5 days (95% CI: 10,8-20,6) versus 8.5 days (95% CI: 6,8-10,2) in CCHD group. The differences are statistically significant – $p < 0.001$ (Figure 2).

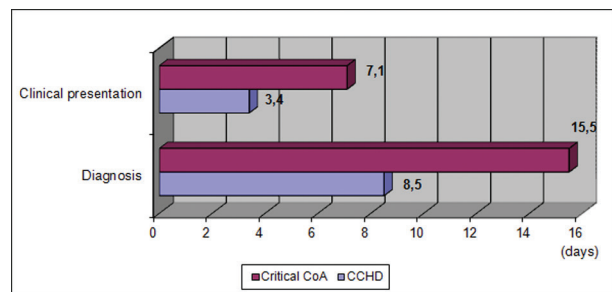


Fig. 2. Time of initial clinical presentation and diagnosis of critical CoA and CCHD

Most of our patients with critical CoA were diagnosed in pediatric departments (PD) – 30 (71.4%). Significantly more often, compared with other CCHD, the diagnosis was established out of

hospitals – 4.8% versus 0.4% and significantly more rare in the neonatology departments (ND) – 9.5%, versus 32.7% ($p < 0,05$) (Figure 3).

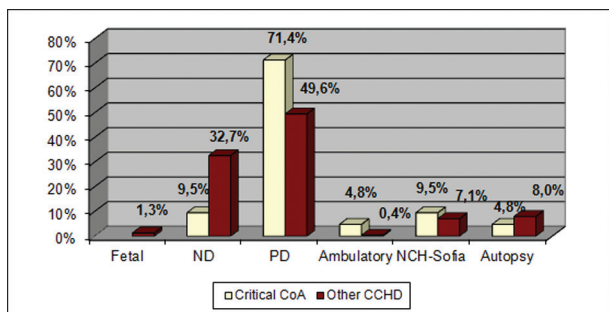


Fig. 3. Place of diagnosing patients with CoA and other CCHD

Fourteen (32.4%) neonates with CoA were dismissed from neonatology departments without cardiac disease, versus 15.8% for CCHD group ($p < 0,05$). In one third of the dismissed critical cardiopathies is CoA (Figure 4).

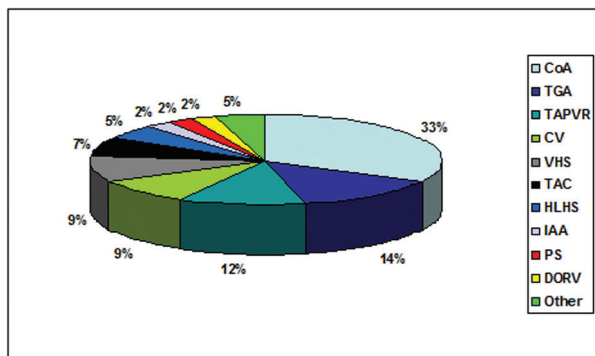


Fig. 4. Newborns with CCHD discharged from neonatology department without being diagnosed

Incorrect or incomplete initial diagnosis was made in 11 of the children (25.6%) against 22% of the whole group with CCHD, without a significant difference ($p > 0,05$).

Corrective procedures were performed in 31 (72.1%) of the patients with CoA – overall 51 interventions, 1.6 on each patient. The presence and the

degree of the concomitant heart malformations were taken under considerations in surgery corrections. Some of the children with hemodynamically isolated CoA - 20 (64.5%) have undergone one-stage procedure and some of them - 10 (32%) - two-stage corrective procedure (banding of the PA and on the second stage – complete correction). One newborn had undergone a Norwood procedure because of hypoplastic ascending aorta (Figure 5). The most frequent operation of the aorta, that had been done in 22 (71%) of the children, is Amato procedure – resection of the aorta and anastomosis “end-to-end”.

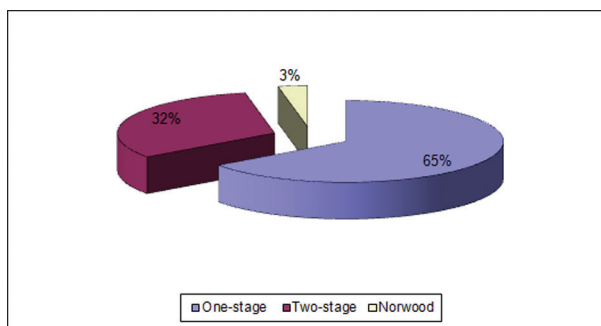


Fig. 5. Types of corrective procedures in critical CoA

Early postoperative mortality rate was 9.7% (3 children), at a mean age of 14.3 days. These newborns were with complex anatomy and concomitant malformations such as Shon’s syndrome, hypoplastic of the aortic arch on a long extend (in one - hypoplastic ascending aorta) and a large VSD.

The mean time of postoperative follow-up was average 6 years. Reoperations were performed in 6 children – 19.4%. Postoperative results were classified as good and very good, altogether 17 (65.4%). Data about two of the children is missing (Table 2).

DISCUSSION

According to our results CoA is the second most frequent CCHD in the region of North-East Bulgaria – 16%. We have established that the clinical manifestation and the diagnosis of the critical

Table 2. Surgical results in CoA

Operative results	Very good	Good	Satisfactory	Unsatisfactory	Unclear	All
Operated with CoA	7	10	6	3	2	28

CoA are significantly delayed in comparison to other CCHD ($p < 0.001$). CoA may form postnatally as a result of the closure of the arterial duct (21,24,28). According to Head CE and colleagues (2005) fetal diagnosis is difficult and post-natal manifestation might be delayed with several weeks (6-12 weeks) and that is the reason serial echocardiography examinations are necessary (15). Our results coincide with the results from other research, showing that the diagnosis is delayed most frequently in newborns with obstructive lesions of the systemic circulation (8) or the cases with CoA, IAA, and TAPVR (31).

Most of our patients were diagnosed in pediatric departments – 71.4% and only 9.5% in neonatology departments, against 32.7% in other CCHD. Comparatively small part of the cases (4.8%), were diagnosed on autopsy. Investigations from Chang and colleagues (2008) from California showed that out of 898 deceased children with CCHD at age from 1 to 364 days, 152 (16.9%) were undiagnosed, mostly with HLHS and CoA, while still living. According to Kearney (2011) about 10% of duct-dependant CCHD remain undiagnosed until death (16).

In neonatology department 14 (32.6%) of the newborns with CoA were undiagnosed, against 15.8% of the all CCHD ($p < 0.05$). One third of the patients with CCHD had critical CoA. According to the research of Mellander and colleagues, 20% of newborns with CCHD, out of 259 patients, who had undergone interventional procedures till the age of two months, were discharged from neonatology department without being diagnosed. The diagnosis was retarded significantly more often in infants with duct-dependant systemic circulations – 30% and duct-independent cardiomyopathies (38%) in comparison to duct-dependant pulmonary circulation – only 4% ($p < 0.001$). The authors specify also a significant increase during the years (1993-2001) in the count of newborns, discharged from hospital without being diagnosed – from 13% to 26% ($p < 0.05$) (19). According to Wren et al. (2007) every third child with life-threatening CHD is discharged from hospital undiagnosed and the most common malformations are CoA – 54%, IAA – 44%, TAPVR – 37% (31). Aamir et al. (2007) also determine that the diagnosis is most often delayed in patients with CoA and the time of the final diagnosis is between the third day and the 6.5th month (1). According to Mouledoux and Walsh

(2013) before the introduction of pulse oximetry, 75% of the cases with undiagnosed CCHD were CoA and it seems that the CoA is the most common missed lesion during routine screening (20).

Incorrect or incomplete diagnosis was made in 11 (25.6%) of the children with CoA, against 22% of the whole group with CCHD. Ainsworth and colleagues (1999) research determine double-incorrect initial diagnoses – 44%, mainly in patients with complex cardiomyopathies, obstructive lesions of the left side of the heart and TAPVR (4).

Kovacikova et al. (2007) indicate that in 34% of the patients with left-sided obstructive lesions, the diagnosis was specified in a transitional medical institution (17). It is recommended a careful cardiac examination, mainly for obstructive lesions of the left-sided heart during the first visit after discharged, between the third and the 5th day of life (8). A number of CHD, especially these with obstructive lesions of the left ventricle outflow tract, may not have a clinical manifestation before the second physical examination and may end up fatally, if left unidentified (2,3,8). The current guidance recommend the clinical screening for CHD in newborns to be followed up by a physical examination after discharged from hospital between the 6th and 8th week of life (9, 14). According to Schultz et al. (2008) most of the “significant physiologic complications”, such as severe metabolic acidosis, convulsions, cardiac arrest or laboratory data for renal or hepatic dysfunction, are presented in unidentified CHD after the first 12 hours – 83%, and 90.9% of the avoidable ones are in obstruction of the aortic arch. According to authors every “reliable” screening should be sensitive for malformations of the aortic arch (25). Pulse oximetry screening has lower sensitivity about left-sided obstructive lesions (18). Sensibility of the fetal echocardiography is lower for left-sided obstructive malformations – 23%, TGA – 19%, TAPVR – 0% (13). Kovacikova et al. also report that prenatal diagnosis has lower sensitivity about the obstructive lesions of the left-side of the heart – in 11% of the cases (17). Das et al. (2012) recommend examination of the natriuretic peptide type B for early identification and treatment of newborns with obstructive lesions of the systemic circulations (11).

In spite of the high frequency of concomitant heart malformations in infantile CoA, also in our

series of operated patients, usually it is possible one-stage resection together with plastic of the aortic arch to be performed – 64.5%. Often, as other authors also say, if there is a concomitant hypoplasia of other left-sided heart structures, such as mitral or aortic valve, it can be overcome after surgical removal of the coarctation and there is no need of additional interventions (22). A preferable operational technique for elimination of coarctation in our patients was Amato procedure - 83%, which is shared by most of the authors (6, 10, 30). In Waldhausen procedure for correction and plastic of the aortic arch in infants with CoA till one year of age, Rafiquil and auth. (1998) report about early post operative hospital lethality – 22,5% (23). Early post operative lethality in our group of patients was 9.7%, reoperations were 19.4% and the distant results were described mostly as good and very good. Perioperative mortality rate, according to the literature data is from 0% till 8.5% and it is significantly higher in patients with heart failure and cardiogenic shock, presented before the corrective surgery (10,12,27,29,30). The most common complications follow surgical correction of CoA were recoarctation – 3-4% and residual hypertension in 25-38% (6, 26).

CONCLUSION

CoA is the second most frequent CCHD in the region of North-East Bulgaria – 16% and the time of initial clinical presentation and diagnosis are significantly delayed ($p < 0.001$). Only in 9.5% of our patients the diagnosis was made in the neonatology department, versus 32.7% in other critical cardiopathies ($p < 0.001$). Newborns with CoA are one third of discharged ones without ascertain diagnosis of cardiac malformation. In one fourth of the children the initial diagnosis was incorrect or incomplete. In most of the patients (64.5%) it was possible a one-stage procedure and major part of the concomitant heart malformations can be reversible. The preferable surgical technique is plastic “end-to-end” by Amato. Early postoperative mortality rate is 9.7%, equally to the literature data.

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