Prevalence of aortic coarctation in Bogotá-Colombia from 2001 to 2018. The diagnostic needs to improve

Prevalencia de coartación aórtica en Bogotá-Colombia de 2001 a 2018. El diagnóstico debe mejorar

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Abstract

Objectives: Aortic coarctation is the most frequent structural anomaly out of congenital heart diseases. This congenital defect is an important cause of death worldwide. We sought to determine the prevalence of aortic coarctation in Colombia and whether new policies have had an impact on its diagnosis. Methods: In this study information from the Bogotá birth defect surveillance program during the years 2001-2018 from 63 hospitals was used. 537,026 live births of any weight and stillbirths of any weight were analyzed. The information was stored in a database on the servers of the Health Secretariat and the Pontificia Universidad Javeriana. We analyzed the presence of aortic coarctation according to the newborn’s sex, weight, size, mother’s age, and gestational age at the time of birth and when coarctation is accompanied by other types of congenital malformations. Results: The prevalence of aortic coarctation in Bogotá during the years 2001-2018 found in this study was 1.25 in 10,000 live births. We also found that prevalence of aortic coarctation in Bogotá changes throughout the years having a significant increase in the year 2018 with 6.57 cases in 10,000 live births. Conclusions: This prevalence is higher than the one found in a study with data from 2001 to 2014, which suggests an improvement in the country’s epidemiological surveillance and medical training. However, the prevalence found in Bogotá is still lower compared to the prevalence worldwide and from other continents, the prevalence for Latinamerica was significantly lower as compared to those in Asia, Europe, and United States so we emphasize the importance of continuing with improvements, such as standardizing screening methods and sensitivity of said methods in a local scale as well as a continental scale.

Keywords: Aortic coarctation. Prevalence. Bogotá. Low birth weight. Gestational age.

Resumen

Objetivos: La coartación aórtica es la anomalía estructural más frecuente de las cardiopatías congénitas. Este defecto congénito es una causa importante de muerte en todo el mundo. Buscamos determinar la prevalencia de la coartación aórtica en Colombia y si las nuevas políticas han tenido un impacto en su diagnóstico. Métodos: En este estudio se utilizó información del programa de vigilancia de defectos de nacimiento de Bogotá durante los años 2001-
2018 en 63 hospitales. Se analizaron 537,026 nacidos vivos de cualquier peso y nacidos muertos de cualquier peso. La información fue almacenada en una base de datos de los servicios de la Secretaría de Salud y de la Pontificia Universidad Javeriana. Se analizó la presencia de coartación aórtica de acuerdo al sexo del recién nacido, peso, tamaño, edad de la madre y edad gestacional en el momento del nacimiento y cuando la coartación se acompañó de otros tipos de malformaciones congénitas. Resultados: La prevalencia de la coartación aórtica en Bogotá durante los años 2001-2018 encontrados en este estudio fue de 1.25 en 10,000 nacidos vivos. También encontramos que la prevalencia de coartación aórtica en Bogotá ha cambiado a lo largo de los años, teniendo un aumento significativo en el año 2018 con 6.57 casos en 10,000 nacidos vivos. Conclusiones: Esta prevalencia es mayor que la encontrada en un estudio con datos de 2001 a 2014, lo que sugiere una mejora en la vigilancia epidemiológica y la formación médica del país. Si bien, la prevalencia encontrada en Bogotá es menor en comparación con la prevalencia a nivel mundial y de otros continentes, la prevalencia para el continente de Latinoamérica también es significativamente menor con respecto a Asia, Europa y Estados Unidos, por lo que enfatizamos la importancia de continuar con las mejoras, como la estandarización de los métodos de detección y la sensibilidad de dichos métodos tanto a nivel local como a nivel del continente.

Palabras clave: Coartación aórtica. Prevalencia. Bogotá. Bajo peso al nacer. Edad gestacional.

Introduction

Aortic coarctation is a type of congenital heart disease. It is the structural anomaly that occurs most frequently. It is classified as an obstructive cardiopathy that takes place in the aorta where it is presented as a narrowing of the diameter at the height of the aortic arch. Depending on the place where the diameter reduction is located, it is given a one-off name: infant or adult. The former consists of a tubular hypoplasia at the aortic arch proximate to the permeable ductus arteriosus. As for the latter, an isolated refold is presented just in front of the arteriosus ligament. The severity of the coarctation can vary and can appear as a single or associated defect to another one, such as aortic stenosis, bicuspid aortic valve, interauricular communication (IAC), interventricular communication (IVC), mitral insufficiency, or even an aneurysm at the Willis polygon.

Worldwide, the prevalence of congenital heart disease is 80 cases/10,000 newborns, 6-8% of these cases have aortic coarctation. In Latin America, 54,000 cases of congenital heart disease are born each year. According to the ECLAMC, the prevalence is 2.6/10,000. In Colombia, according to the Ministry of Health, the prevalence of congenital heart disease was 75-95/10,000 newborns in 2006. In Bogotá, according to a study carried out during the years 2001 and 2014, the prevalence of aortic coarctation is 0.7/10,000 newborns.

Clinical manifestations depend on the degree of tightness and permeability of the ductus arteriosus. Due to the reduced diameter of the aorta systolic arterial hypertension is common, presenting in up to one-third of patients. Moreover, aortic coarctation with a patent ductus arteriosus (PDA) can manifest itself in the early stages of life. Many newborns do not survive the neonatal period if no surgery is done. In cases where an intervention is not performed cyanosis will result in the lower limbs as a result of unsaturated blood. On the other hand, aortic coarctation with no persistent ductus arteriosus can be asymptomatic in children and they can even reach adulthood without being identified.

Characteristic findings include patients with hypertension in the upper limbs and hypotension in the lower limbs. There are also symptoms such as claudication and coldness in the lower limbs. In adults with aortic coarctation, markings can be found on their ribs. Such phenomenon is attributed to the presence of collateral irrigation coming from the intercostal and internal mammary arteries. Systolic murmurs along with the left ventricular hypertrophy due to afterload increase may be found.

The best way to screen patients for congenital heart disease is pulse oximetry 24-48 h after birth. In Colombia, the latter appeared for the 1st time in a clinical guideline in the year 2013; however, it has been implemented since 2018 due to government policies. The procedure must be done on the right hand and on any foot. The latter is done in such a manner because the right hand provides preductal saturation, whereas any foot provides a postductal saturation. The difference between preductal and postductal saturation in a newborn without congenital heart disease should be ≤ 3% and saturation in either extremity should be greater than 95%. In this case, normal nursing of the newborn is performed. However, other congenital heart
diseases are not completely ruled out and other findings in the physical examination must be taken into account.

In another scenario, when pulse oximetry is positive the saturation is <90% in the right limb or foot. An echocardiogram should be requested and the causes of hypoxemia should be sought. It can also test positive with a saturation between 90% and 94% and a difference greater than 4%. Given the latter scenario, the oximetry must be repeated within 1 h. If negative, normal nursing of the newborn is performed and if positive then the test is repeated within 1 h and an echocardiogram is requested and other causes of hypoxemia are sought\textsuperscript{10,11}.

It is important to mention that using pulse oximetry as a screening method has a sensitivity of 80% and that false negative oximeter results are not rare. This may be due to small right-to-left shunt or a relatively high cardiac output and mixed venous saturation. Such findings are more common in pathologies such as coarctation of the aorta\textsuperscript{12}.

Diagnosis can be made before or after the baby is born. Prenatal diagnosis is difficult mainly because the presence of the ductus arteriosus makes diagnosis unclear. Coarctation is more visible during weeks 16-18 of gestation and is generally diagnosed when accompanied by more complex heart conditions. On the other hand, postnatal diagnosis made when systolic hypertension in the upper limbs and delayed femoral pulses are present. The latter is confirmed by an echocardiography\textsuperscript{2,8,13}.

**Methods**

The information recorded in the Bogotá birth defect surveillance program for 537,026 live births of any weight and stillbirths of any weight was analyzed. All births between January 2, 2000 and June 30, 2018, at 63 hospitals were included in the study. The surveillance program used is a monitoring and tracking program of children with congenital defects that uses the information provided on the National Health Surveillance System (Sivigila) by all hospitals in Bogotá and registers the cases by active surveillance at seven sentinel hospitals in the city.

The information was coded according to the ICD-10/ British Pediatric Association and was stored in a database on the servers of the Health Secretariat and the Pontificia Universidad Javeriana. Newborns in the seven sentinel hospitals were evaluated by a physician trained in the detection of congenital malformations, considering as case the child having a congenital malformation as described in the ECLAMC operational manual. For this evaluation, the signature of informed consent and the endorsement of the Ethics and Research Committee of the Pontificia Universidad Javeriana and Hospital Universitario San Ignacio were requested. In the other hospitals, the congenital defect sheet was filed according to the protocol published on Colombia’s National Health Institute website (www.ins.gov.co).

The following variables were analyzed: the distribution of congenital heart disease “coarctation of the aorta” (ICD-10/BPA code: Q25.19) according to the newborn’s sex, weight, size, the age ranges of the mother, the gestational age at the time of birth, and the frequency with which coarctation is accompanied by other type of congenital malformations. Collected information was recorded in a modified version of the same database in the Microsoft Excel program. It was evaluated whether the aforementioned variables were or were not risk factors for presenting aortic coarctation.

Isolated coarctation was defined as the one which was not associated with any other congenital malformation. Complex cardiopathy was defined as the one in which two or more cardiac abnormalities were present. Polymalformed cases were defined as those which presented one or more non-cardiac malformations. Classification of syndromics, among those contemplating trisomy 18, trisomy 21, Klippel Feil syndrome, and Goldenhar syndrome, were also included. The prevalence was calculated by taking the total number of children with aortic coarctation and using the number of total newborns as denominator.

**Data analysis**

Quantitative variables were compared using t-tests with a 95% confidence interval (CI). Qualitative variables were analyzed using Odds ratio with a 95% CI. Microsoft Excel for Mac version 16.34 and Epicalc 2000 v1.0.2.1 were used for analyzing the information and statistics calculation, respectively.

**Results**

During the years 2001 and 2018, 67 aortic coarctation cases were identified in 537,026 births. The prevalence found was 1.25 in 10,000 live births. The collected data come from 22 hospitals in Bogotá, Colombia.
Table 1. Distribution of congenital malformations associated with aortic coarctation, according to isolated cases, complex cardiopathies, polymalformed, or syndromics

| Malformation                                                                 | ICD-10/BPA code | Cases | Percentage of total cases |
|------------------------------------------------------------------------------|-----------------|-------|--------------------------|
| Isolated coarctation                                                         |                 | 30    | 44.8%                    |
| Complex cardiopathy                                                          |                 |       |                          |
| Interauricular communication                                                  | Q21.1           | 27    | 40.3%                    |
| Interventricular communication                                                | Q21.0           | 14    |                          |
| PDA                                                                          | Q25.0           | 11    |                          |
| Hypoplastic left heart syndrome                                               | Q23.4           | 8     |                          |
| Double inlet left ventricle                                                  | Q20.4           | 5     |                          |
| Other congenital malformations of aorta                                       | Q25.4           | 3     |                          |
| Congenital mitral stenosis                                                   | Q23.2           | 3     |                          |
| Other congenital malformations of pulmonary valve                            | Q22.3           | 3     |                          |
| Other congenital malformations of other great arteries                       | Q25.8           | 3     |                          |
| Other specified congenital malformations of heart                            | Q24.8           | 2     |                          |
| Discordant ventriculoarterial connection                                      | Q20.3           | 2     |                          |
| Other congenital malformations of tricuspid valve                            | Q22.8           | 1     |                          |
| Congenital absence and hypoplasia of umbilical artery                        | Q27.0           | 1     |                          |
| Polymalformed cases                                                          |                 | 6*    | 9.0%                     |
| Congenital absence and hypoplasia of umbilical artery                        | Q27.0           | 3     |                          |
| Congenital absence, atresia and stenosis of anus with fistula                | Q42.2           | 2     |                          |
| Polydactyly                                                                  | Q69             | 2     |                          |
| Multiple congenital malformations, not elsewhere classified                  | Q89.7           | 1     |                          |
| Hypospadias balanic                                                          | Q54.0           | 1     |                          |
| Phocomelia, unspecified limb                                                  | Q73.1           | 1     |                          |
| Micrognathia                                                                 | Q67.4           | 1     |                          |
| Congenital diaphragmatic hernia                                              | Q79.0           | 1     |                          |
| Undescended testicle, unspecified                                           | Q53.9           | 1     |                          |
| Other congenital malformations of pulmonary valve                            | Q22.3           | 1     |                          |
| PDA                                                                          | Q25.0           | 1     |                          |
| Syndromics                                                                   |                 | 4     | 6.0%                     |
| Trisomy 18                                                                   | Q91.0           | 1     |                          |
| Trisomy 21                                                                   | Q90.0           | 1     |                          |
| Klippel-Feil syndrome                                                        | Q76.1           | 1     |                          |
| Goldenhar syndrome                                                           | Q87.0           | 1     |                          |
| Total                                                                        |                 | 67    | 100%                     |

*The sum of malformations below exceeds the total number of cases in this category, since some patient’s present more than one congenital malformation besides coarctation. ICD: International Classification of Diseases; PDA: patent ductus arteriosus.

As shown in Table 1, from the 67 coarctation cases, 30 cases presented as isolated coarctation and 27 as complex cardiopathy predominantly accompanied by IAC, IVC, and PDA. Six presented as polymalformed cases accompanied by single umbilical artery, imperforate anus, and other types of polydactylism. Finally, four cases presented as syndromics.

Mean weight, mean height, mean maternal age, and mean gestational age were calculated for the group of coarctation cases and all births during the years 2001-2018, finding the results are described in Table 2.

We found that the prevalence of aortic coarctation in Bogotá changes in different years, generating a multimodal graph with a significant increase in the year 2018 with 6.57 cases in 10,000 live births as shown in figure 1.

Discussion

Prevalence data worldwide

Congenital cardiopathies are the leading cause of birth defects and their prevalence has changed dramatically and geographically over time. These changes may be associated with an under diagnosis since the lowest prevalence was found in Africa and Latin America and the highest in Asia, Europe, and the United States. Aortic coarctation is the sixth congenital cardiopathy with the highest prevalence, which was 3.4/10,000 live births worldwide. In a systematic review and meta-analysis which analyzed prevalence of congenital cardiopathies between 1970 and 2017, 260 studies were included and the period with the highest...
The prevalence was during the years 2010-2017 with a prevalence of 94.1/10,000 live births. This is explained by the improvement in postnatal diagnosis of these malformations as a result of implementation of better quality echocardiographic equipment and training of clinical personnel. The calculated prevalence of coarctation of the aorta was 2.87/10,000 live births, similar to other prevalence’s found in systematic reviews with data from different populations\textsuperscript{15}. This strengthens our theory that there might be an under-registration and thus an under-diagnosis of this particular malformation throughout the continent.

It should be noted that the early diagnosis of congenital heart diseases, such as aortic coarctation, also depends on having a good surveillance system. It is known that some programs as the National Network of Congenital Anomalies of Argentina (RENAC), Congenital Disease Registry Center of Costa Rica and the Epidemiological Surveillance System for Neural Tube Defects and Craniofacial Defects in Mexico, monitor newborns even 1 year after they are born, which not only improves surveillance of the population allowing to report congenital anomalies not detected at the time of birth and thus having more reliable prevalence rates, but also allows to make scientific publications about the congenital anomalies in their territories. For instance, in Argentina and Costa Rica, where their surveillance systems share data with other international entities such as the International Clearinghouse for Birth Defects Surveillance and Research\textsuperscript{17}. With this information, we can say that Colombia currently has a congenital heart disease screening program and a high-quality surveillance system for congenital defects, in which the aforementioned monitoring is also carried out, compared to other Latin American countries, which has made it possible to improve national indicators in the past few years.

### Prevalence data in Europe (EUROCAT)

The ECLAMC (Latin American Collaborative Study of Congenital Malformations) is a clinical and epidemiologic research program to which cases of congenital malformations are reported in Latin America. Analyzing data between the years 1995 and 2008, the prevalence of congenital cardiopathies in Latin America is of 35.3/10,000 live births; however, said data do not specify prevalence of aortic coarctation\textsuperscript{16}. This strengthens our theory that there might be an under-registration and thus an under-diagnosis of this particular malformation throughout the continent.

Other studies in Bogotá

In a study that took place in Bogotá between the years 2001 and 2014\textsuperscript{3}, to determine the prevalence of

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**Table 2.** Quantitative variable comparison between coarctation cases and all births during the years 2001-2018

| Variable          | Cases 2001-2018 (n = 67) | All births during 2001-2018 (n = 536,959) | p-value |
|-------------------|--------------------------|------------------------------------------|---------|
| Birth weight (g)  | 2,828.78±621.21          | 2,992.1±522.43                          | 0.018*  |
| Size (cm)         | 48.2±3.25                | 49.42±2.97                              | 0.001*  |
| Maternal age (y)  | 27.51±6.43               | 26.25±6.48                              | 0.219   |
| Gestational age (wk) | 37.77±2.15             | 38.26±2.11                              | 0.035*  |

*p < 0.05); SD: standard deviation.

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**Figure 1.** Prevalence of aortic coarctation in Bogotá by years per 10,000 births.
congenital cardiopathies information from 405,408 newborns registered in the Surveillance and Follow-up Program for Children with Congenital Anomalies of the Health Secretary of Bogotá was analyzed. The prevalence calculated for congenital cardiopathies was of 15.1/10,000 live births. From the children born with malformations, 397 were classified as isolated, 142 were associated with extracardiac malformations and 74 as complex cardiopathies. The prevalence found for aortic coarctation in this study was of 0.7/10,000 live births. This is a smaller prevalence than the one found in the present study which was of 1.25/10,000 live births.

In comparison with a different study performed in Bogotá that studied prevalence of congenital cardiopathies in a neonatal unit hospital in Bogotá between the years 2011 and 2015 the prevalence for aortic coarctation was 2.39/10,000 live births, which is also different from the prevalence we found in our current study. The difference between these prevalence’s can be associated with the level of complexity of the hospital from which the data were collected. The study between the years 2011 and 2015 took data from a high complexity hospital that has a neonatal unit, to which most critical patients are referred to, and these patients are more likely to present congenital cardiopathies like aortic coarctation. On the contrary, the data taken for our current study come from 63 different hospitals in Bogotá from a variety of complexity levels. This is why some of those hospitals may have an under registration of these cardiopathies.

Associated variables

In our study, we found an association between aortic coarctation and low birth weight according to gestational age, as well as low birth height, with data from the World Health Organization. Compared with a study conducted in Houston, Texas, in which growth patterns in infants with cardiovascular malformations were studied, it was found that our results match. In this study, they came to the conclusion that children with aortic coarctation were at higher risk of presenting low birth weight and low birth height compared with births in the same period.

When analyzing the average maternal age between cases and births from the same time period, no significant difference is evidenced. This matches with the findings of a study conducted in the United States that took data between the years 1999 and 2007 in which the prevalence of different cardiovascular malformations was analyzed by dividing the maternal ages in different 5-year periods. This study concluded that there is no association between maternal age and aortic coarctation. The same study also showed an increase in the prevalence of aortic coarctation in 17% between the years 1999-2001 and 2005-2007. In our current study, the year 2007 was the 2nd year with the highest prevalence (2.87/10,000 live births) followed by the year 2018 (6.56/10,000 live births). It was in 2018 when we found a peak in prevalence of aortic coarctation, as shown in Graph 1, which can be explained due to an increase in detection of this pathology attributed to enhancements in the registration and follow-up of the cases.

The increase of cases in the year 2018 may also be the result of the emphasis that health authorities have placed on the detection of congenital cardiopathies; taking into account that these congenital defects have the highest impact on child mortality and therefore registration must have increased. The low prevalence, compared with Europe or the United States, may also be due to the approval of voluntary termination of pregnancy since the year 2006, and consequently some cases are not carried to full-term after prenatal diagnosis.

Conclusion

The current study found a higher prevalence for aortic coarctation in Bogotá in comparison with the study conducted by Tassinari et al. that worked with the same surveillance program: Surveillance and Follow-up Program for Children with Congenital Anomalies of the Health Secretary of Bogotá. Tassinari’s et al. work mentioned the need to “strengthen epidemiologic surveillance programs in the country [...] and to improve medical training from the most basic levels of healthcare to facilitate detection of congenital abnormalities.”

Taking into account that the study conducted by Tassinari et al. took data during the years 2001-2014 and our study included data up to the year 2018, the increase in prevalence may suggest that improvements were made according to the authors’ proposals during said range of time. However, the prevalence found in Bogotá is still lower compared to prevalence worldwide and in other continents. Furthermore, the prevalence found in Latinamerica was significantly lower compared to prevalence in Asia, Europe, and The United States.

As a result of the information previously mentioned, we emphasize the importance of standardizing screening techniques in a local scale, as well as in a continental scale, so that every hospital can apply them, and no newborn is discharged without a diagnosis and a
proper notification. Furthermore, it is important to intervene in factors that might decrease prevalence, such as low sensitivity of prenatal and postnatal diagnosis to make the statistics more accurate.

Colombia currently has a high quality congenital cardiopathy screening program as well as a surveillance system for congenital malformations in comparison to other countries in Latin America. This has allowed us to improve national statistics regarding diagnosis, thus we recommend that the health personnel from other countries in the continent review this experience to improve healthcare quality.

Disclaimer

All of our variables were selected, including maternal age, which was selected to confirm that it had no significance. Taking into account the database of births of the Health Secretariat, from which we took the data for our article, it is not possible to follow the evolution of patients. Furthermore, the analysis of treatments and their outcome is beyond the scope and objective of this article.

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Conflicts of interest

None.

Ethical disclosures

Protection of human and animal subjects. The authors declare that no experiments were performed on humans or animals for this study.

Confidentiality of data. The authors declare that they have followed the protocols of their work center on the publication of patient data.

Right to privacy and informed consent. The authors have obtained the written informed consent of the patients or subjects mentioned in the article. The corresponding author is in possession of this document.

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