rfm
Revista de la Facultad de Medicina

ORIGINAL RESEARCH

Incidence and geographic distribution of biliary atresia in children under 1 year of age in Colombia during the period 2018-2021

Incidencia y distribución geográfica de la atresia biliar en Colombia en menores de 1 año durante el periodo 2018-2021

Angie Vanessa Vergara-Espitia[1] John Jairo Zuleta-Tobón[2] Patricia Ruiz-Navas[3] Margarita María Suarez-Galvis[3]

[1] Universidad El Bosque - Faculty of Medicine - Specialty in Pediatric Gastroenterology - Bogotá D.C. - Colombia.
[2] Hospital Pablo Tobón Uribe - Department of Epidemiology - Medellín - Colombia.
[3] Hospital Pablo Tobón Uribe - Liver Transplantation Group and Inborn Errors of Metabolism Group - Medellín - Colombia.

Open access

Received: 14/02/2024
Accepted: 23/09/2024

Corresponding author: Angie Vanessa Vergara-Espitia. Especialización en Gastroenterología Pediátrica, Facultad de Medicina, Universidad El Bosque. Bogotá D.C. Colombia. E-mail: angievve1@gmail.com.

Keywords: Biliary Atresia; Children; Cholestasis; Epidemiology; Liver transplantation (MeSH).

Palabras clave: Atresia biliar; Niños; Colestasis; Epidemiología; Trasplante hepático (DeCS).

How to cite: Vergara-Espitia AV, Zuleta-Tobón JJ, Ruiz-Navas P, Suarez-Galvis MM. Incidence and geographic distribution of biliary atresia in children under 1 year of age in Colombia during the period 2018-2021. Rev. Fac. Med. 2024;72(4):e112968. English. doi: https://doi.org/10.15446/revfacmed.v72n4.112968.

Cómo citar: Vergara-Espitia AV, Zuleta-Tobón JJ, Ruiz-Navas P, Suarez-Galvis MM. [Incidencia y distribución geográfica de la atresia biliar en Colombia en menores de 1 año durante el periodo 2018-2021]. Rev. Fac. Med. 2024;72(4):e112968. English. doi: https://doi.org/10.15446/revfacmed.v72n4.112968.

Abstract

Introduction: Biliary atresia (BA) is the most common cause of obstructive jaundice in the first months of life. If not treated promptly, BA rapidly progresses to biliary cirrhosis and eventually leads to early death.
Objective: To estimate the incidence of BA in children under 1 year of age in Colombia and its territorial divisions (32 departments and one special district) for the period 2018-2021.
Materials and methods: Cross-sectional study using secondary data. Cases of BA in children under 1 year of age officially reported in Colombia to the National Institute of Health (INS by its Spanish acronym) between 2018 and 2021 were analyzed. The annual incidence of BA was calculated by dividing the number of BA cases reported each year for children under 1 year of age at the time of notification by the number of live births (LBs) in that same year. Incidence was also calculated at the national level and in each of the country's territorial divisions (32 departments and 1 special district) for the period 2018-2021.
Results: Between 2018 and 2021, 82 cases of BA were reported in children under 1 year of age in Colombia, of which 58.54% involved girls. The median age at the time of notification was 4.89 months. The incidence of BA in Colombia for the period 2018-2021, based on the number of cases reported for children under 1 year of age at the time of official notification of the event to the INS, was 3.23 cases per 100 000 LBs (annual range: 2.19 and 4.05 cases). The departments of Boyacá, Santander and Cauca had the highest incidence rates (7.5, 7.212 and 6.374 cases per 100 000 live births), while the departments of Magdalena, Córdoba and Bolívar had the lowest (0.747, 0.98 and 1.02 per 100 000 live births).
Conclusions: The incidence of BA in Colombia for the period 2018-2021, based on the number of cases in which the age of notification was under 1 year, was 3.23 cases per 100 000 LBs, which is lower than what has been reported in other countries and regions.

Resumen

Introducción. La atresia biliar (AB) es la causa más común de ictericia obstructiva en los primeros meses de vida. Si no se trata de forma oportuna, la AB progresa rápidamente a cirrosis biliar y eventualmente lleva a la muerte temprana.
Objetivo. Estimar la incidencia de la AB en menores de 1 año en Colombia y en sus divisiones territoriales (32 departamentos y un distrito especial) para el periodo 2018-2021.
Materiales y métodos. Estudio transversal realizado con datos de fuente secundaria. Se analizaron los casos de AB en menores de 1 año notificados oficialmente en Colombia ante el Instituto Nacional de Salud entre 2018 y 2021. La incidencia anual de AB se calculó dividiendo el número de casos de AB reportados cada año en niños que eran menores de 1 año al momento de la notificación por el número de recién nacidos vivos (NV) en ese mismo año. También se calculó la incidencia nacional y en cada una de las divisiones territoriales del país (32 departamentos y 1 distrito especial) para el periodo 2018-2021.
Resultados. Entre 2018 y 2021 se notificaron 82 casos de AB en menores de 1 año en Colombia, de los cuales 58.54% eran niñas. La mediana de edad al momento de notificación fue de 4.89 meses. La incidencia de AB en Colombia para el periodo 2018-2021, basada en el número de casos reportados en niños que tenían menos de 1 año al momento de la notificación oficial del evento ante el INS, fue 3.23 casos por cada 100 000 NV (rango anual: 2.19 y 4.05 casos). Los departamentos de Boyacá, Santander y Cauca tuvieron las tasas de incidencia más altas (7.5, 7.212 y 6.374 casos por cada 100 000 NV), mientras que los departamentos de Magdalena, Córdoba y Bolívar, las más bajas (0.747, 0.98 y 1.02 por cada 100 000 NV).
Conclusiones. La incidencia de AB en Colombia para el periodo 2018-2021, basada en el número de casos en los que la edad de notificación fue menor a 1 año, fue de 3.23 casos por cada 100 000 NV, la cual es inferior a la reportada en otros países y regiones.

Introduction

Biliary atresia (BA) is a progressive fibroinflammatory cholangiopathy that affects the intrahepatic and extrahepatic bile ducts, resulting in their partial or total obstruction.[1-3] Its etiology is unknown[4-6] and a timely diagnosis is of great importance since, if left untreated, it can lead to a fatal outcome in the first two years of life.[2,3,6]

BA is considered a surgical emergency and Roux-en-Y portoenterostomy (Kasai procedure) is the gold standard for its treatment.[4-6] However, age is a factor directly related to the effectiveness of this procedure in restoring bile flow and, therefore, improving the survival of the native liver. It has been reported that if this bypass surgery is performed in the optimal time window (first 60 days),[4-6] flow will be restored in about 75% of cases, but if it is performed after 90 days of life, the success rate is less than 25%.[4] Similarly, although it has not been systematically evaluated, the surgeon's expertise in the performance of portoenterostomy has also been associated with the outcome.[4] On the other hand, liver transplantation is the only treatment option in cases in which, due to late detection, advanced liver fibrosis (cirrhosis) is present or when the Kasai procedure is unsuccessful in restoring bile flow.[3,4]

BA is the main cause of neonatal cholestasis,[1,2,4] the most frequent identifiable cause of obstructive jaundice in the first 3 months of life,[4] and the most common cause of end-stage liver disease in the first 2 years of life, being the most frequent indication for liver transplantation in the pediatric population (40-50% of cases).[1-3]

The incidence of BA varies widely both between and within regions and countries.[5-7] For example, in 2013, Jimenez-Rivera *et al.*[5] reported in a systematic review that included 40 studies on BA in the world, with 28 of them reporting data on population-based incidence or prevalence of infants with BA, that the incidence of this condition varies between 0.12 and 3.7 cases per 10 000 live births (LBs); however, that study did not include any data for Colombia (Figure 1).[5] Regarding prevalence, authors such as Fawaz *et al.*[4] describe that the prevalence varies according to location, ranging from 1 in 6 000 LBs to 1 in 19 000 LBs (1 in 6 000 in Taiwan, 1 in 12 000 in the United States, 1 in 18 000 in Europe, and 1 in 19 000 in Canada).

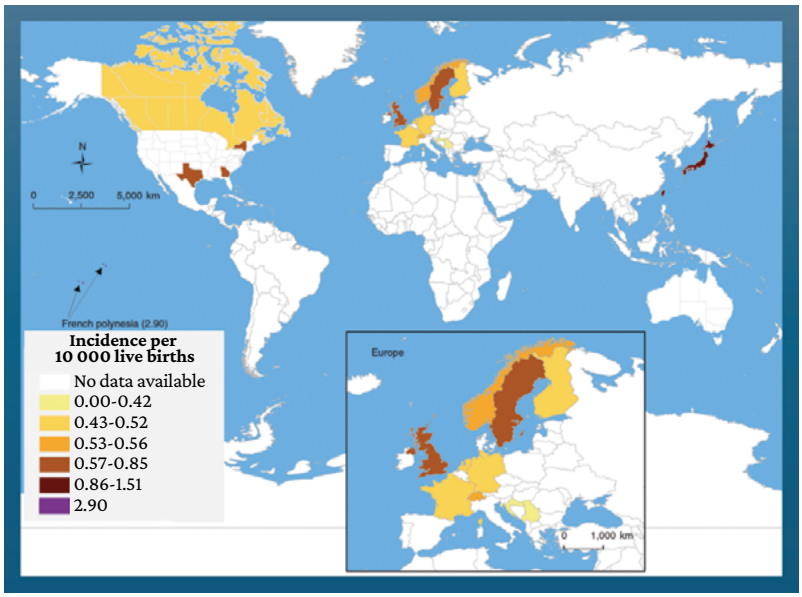


Figure 1. Incidence of infants with biliary atresia worldwide.
Source: Adapted from Jimenez-Rivera *et al.*[5]

Knowing the incidence and geographic distribution of BA for a specific country is of great importance, as this information will allow focusing efforts to improve early diagnosis rates, which, as mentioned above, has a great positive impact on the prognosis of these patients.

Considering the foregoing, the objective of the present study was to estimate the incidence of BA in children under 1 year of age in Colombia and in its territorial divisions (32 departments and one special district) for the period 2018-2021.

Materials and methods

Study type

Cross-sectional study conducted using secondary source data.

Data used

We analyzed data on BA cases officially reported in Colombia between 2018 and 2021. In other words, we analyzed the cases reported to the National Institute of Health (INS by its Spanish acronym) by means of a notification form for this event of health interest, which is of mandatory notification since 2016.[8] It should be noted that there is no time limit for reporting the event, hence we refer to children under 1 year of age at the time of official notification of the event and not to children under 1 year of age at the time of diagnosis. This information was provided by the INS in a database created in Microsoft Excel.

Data analysis

Data are described using absolute frequencies and percentages for qualitative variables and means or medians with their respective standard deviations or 25th and 75th percentiles for quantitative variables according to the distribution of the data.

The annual incidence of BA for the country was calculated by dividing the number of BA cases reported each year for children under 1 year of age at the time of notification by the number of newborns in that same year. Furthermore, the incidence countrywide and in each territorial division (32 departments and one special district) of BA for the period 2018-2021 was also calculated. The year 2022 was not taken into account because it was considered that the report of BA cases in children under 1 year of age for that year was still preliminary at the time the database was obtained. The number of births by year and by department is publicly available in the official birth databases of the National Administrative Department of Statistics (DANE by its Spanish acronym) - Vital Statistics of Births and Deaths.[9] Statistical analyses were performed in the STATA software (version 17.0) and the incidence map in Microsoft Excel.

Ethical considerations

The present study, as an analysis of publicly available secondary data, does not require approval by an ethics committee. However, following the ethical principles of biomedical research, the study was submitted to the Institutional Ethics Committee of the Hospital Pablo Tobón Uribe, obtaining its approval as stated in Minutes No. 05/2023 of March 16, 2023.

Results

Between 2018 and 2021, 82 BA cases were reported in children under 1 year of age in Colombia, of which 58.54% were female. The case with the youngest age at the time of notification was 19 days old, and the median age was 4.89 months (25th percentile: 3 months; 75th percentile: 7 months).

These 82 cases were reported in 20 of the 33 territorial divisions of the country, with the departments of Antioquia (18.29%), Valle del Cauca (14.63%), and the Special District of Bogotá (13.41%) being the divisions with the highest number of reports (Table 1). In addition, 95.12% (n=78) of these children resided in urban areas or municipal seats.

Table 1. BA cases in children under 1 year of age at the time of notification reported in the period 2018-2021. Distribution by department or territorial division.

Department	Reported cases in children under 1 year of age n (%)
Amazonas	0 (0.00)
Antioquia	15 (18.29)
Arauca	0 (0.00)
Atlántico	5 (6.09)
Special District of Bogotá	11 (13.41)
Bolivar	3 (3.65)
Boyacá	4 (4.87)
Caldas	0 (0.00)
Casanare	1 (1.21)
Caquetá	0 (0.00)
Cauca	4 (4.88)
Cesar	4 (4.88)
Choco	1 (1.21)
Córdoba	1 (1.21)
Cundinamarca	2 (2.44)
Guainía	0 (0.00)
Guajira	2 (2.44)
Guaviare	0 (0.00)
Huila	0 (0.00)
Magdalena	2 (2.44)
Meta	2 (2.44)
Nariño	2 (2.44)
Norte de Santander	1 (1.21)
Putumayo	0 (0.00)
Quindío	1 (1.21)
Risaralda	0 (0.00)
Santander	8 (9.75)
San Andrés and Providencia	0 (0.00)
Sucre	1 (1.21)
Tolima	0 (0.00)
Valle del Cauca	12 (14.63)
Vaupés	0 (0.00)
Vichada	0 (0.00)
Total	**82 (100)**

The incidence of BA in children under 1 year of age at the time of event notification for the period 2018-2021 in Colombia was 3.23 cases per 100 000 LBs and varied annually between 2.19 and 4.05 cases per 100 000 LBs (Figures 2 and 3).

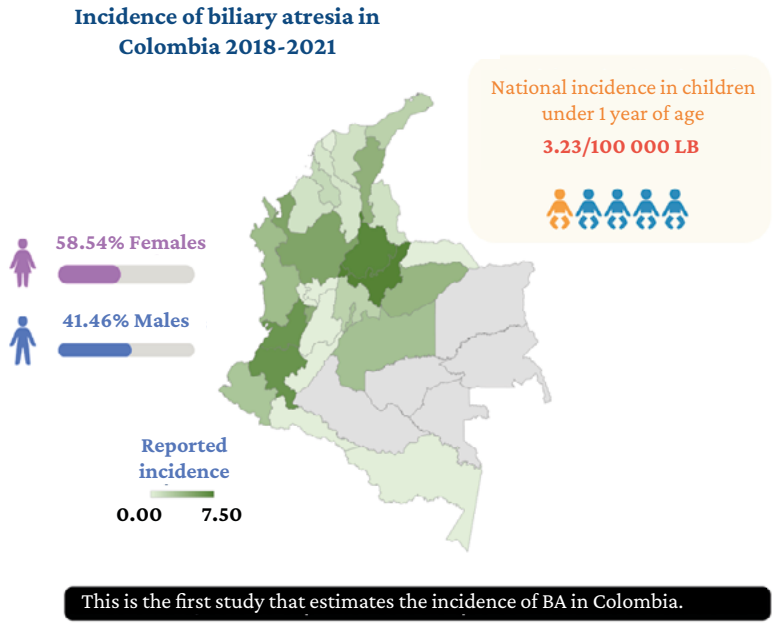
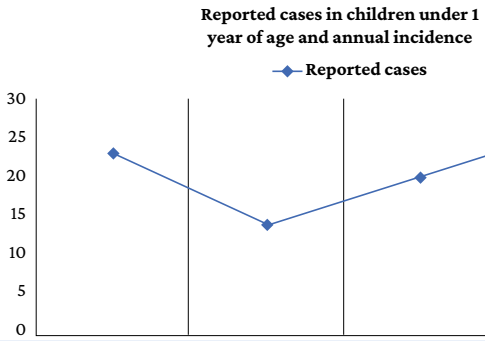


Figure 2. Incidence of biliary atresia in Colombia among children under 1 year of age in the period 2018-2021.



	2018	2019	2010	2011	Total in 4 years
Reported cases	23	14	20	25	82
LB (live births)	649 115	639 820	629 402	616 914	
Incidence	3.54	2.19	3.18	4.05	3.23

Figure 3. Incidence of biliary atresia in Colombia. Annual incidence and for the period 2018-2021.

Regarding the incidence of BA for the period 2018-2021 in the 20 territorial divisions in which BA cases were reported in children under the age of 1 year at the time of the official case report, the following was found: the highest incidence rates were observed in the departments of Santander, Boyacá, and Cauca with 7.21, 7.05, and 6.37 cases per 100 000 LBs, respectively. In contrast, Magdalena, Córdoba, and Bolívar had the lowest incidence rates, with 1.02, 0.98, and 0.74 cases per 100 000 LBs, respectively. In the Special District of Bogotá, the incidence was 3.05 cases per 100 000 LBs (Table 2).

Table 2. Incidence of biliary atresia from 2018-2021 by territorial division in Colombia.

Department	Cases per 100 000 live births
Antioquia	5.19
Barranquilla	5.39
Special District of Bogotá	3.05
Bolívar	0.74
Boyacá	7.05
Cartagena	2.92
Casanare	3.99
Cauca	6.37
Cesar	4.33
Choco	3.56
Córdoba	0.98
Cundinamarca	2.04
Guajira	2.11
Magdalena	1.02
Meta	3.27
Nariño	2.96
Norte de Santander	1.03
Quindío	4.48
Santander	7.21
Special District of Santa Marta	3.07
Sucre	1.57
Valle del Cauca	6.22

Finally, it should be noted that the database provided by the INS only includes the date of notification of this event of health concern (i.e., BA) and not the age at diagnosis. Therefore, it was only possible to determine the age at the time of notification, with an older median age of notification for the 82 cases reported between 2018 and 2021, as previously reported. It was also observed that the reporting of BA cases in which the age of the patient at the time of notification was less than 1 year progressively increased over the study period, rising from 28% in 2018 to 55% in 2021.

Discussion

As mentioned above, the prevalence of BA varies worldwide between 1 in 6 000 LBs and 1 in 19 000 LBs.[4] Similarly, it has been described that its incidence varies widely both between and within regions and countries[5-7] and multiple authors have studied it in different countries.[5] However, there are no studies that report this type of data for Colombia, this being, to our knowledge, the first study that provides information on the subject.

According to the data presented in our study, the incidence rate of BA in Colombia for the period 2018-2021, based on the number of cases reported in children who were under 1 year old at the time of official notification of the event to the INS, was 3.23 cases per 100 000 LBs. This condition has been reported to be much more prevalent in Asian

countries than in Western countries.[1,4,5,7,10] For example, it has been described that the incidence in Korea is 10.6 cases per 100 000 LBs;[7,10] in Japan, 11 cases per 100 000 LBs;[7,10] in Taiwan 14.8 cases per 100 000 LBs;[7,10] in Hong Kong, 12 cases per 100 000 LBs;[7] and in China, 20 cases per 100 000 LBs;[7] whereas in Western countries the incidence varies between 5.2 and 7.1 cases per 100 000 LBs.[7,10] In a literature review conducted to develop a guideline for the evaluation of cholestatic jaundice in infants, Fawaz *et al.*[4] found that an incidence of 5.5 and 5.26 cases per 100 000 LBs has been reported in Europe and Canada, respectively. Also, in a study using data from 1 057 children under 1 year of age who were diagnosed with BA between 1997 and 2012 and had undergone the Kasai procedure, Hopkins *et al.*[6] described an incidence of 4.47 cases per 100 000 children under 1 year of age in the United States, identifying an average annual increase of 7.9% in the incidence of this condition during that period.

Based on the foregoing and on our findings, it is possible to state that the incidence of BA in Colombia is lower than in other countries and regions, although this could be the result of underreporting of this condition in the country.

On the other hand, in the whole group, a predominance of the female sex was observed since, of the 82 cases reported between 2018 and 2021 in the country, 58.54% were girls. This finding is in line with what is described in the literature, in which it is reported that this condition tends to affect the female sex more.[1,3,6] In this regard, Hopkins *et al.*,[6] in a study conducted in the United States, reported that the incidence of BA in the period 1997-2012 was higher in girls than in boys (5.36 vs. 3.74 cases per 100 000 children under 1 year of age; RR: 1.43, 95%CI: 1.27-1.62).

A noteworthy finding is that, while the age at diagnosis could not be determined due to the characteristics of the database analyzed, the age at the time of notification of the BA cases was quite advanced (median age at the time of notification: 4.89 months). Even though it is not possible to establish the time lapse between BA diagnosis and its official notification to the INS, it could be assumed that, despite the fact that there is no time limit for reporting this health event, and considering that reporting this health event is mandatory in the country, this time interval should not be longer than some weeks; therefore, it could be concluded that, in general, the age of diagnosis of BA in the country is late. This delay is detrimental to the prognosis of these patients, as it has been reported that a timely diagnosis (before 45-60 days of life) is essential to optimize the effectiveness of the Kasai procedure to reestablish bile flow and, therefore, improve the survival of the native liver.[1,2,4,6]

At this point, it should be stressed that a late BA diagnosis is a frequent problem worldwide for several reasons, including a visual overlap that is taken for normal physiologic jaundice and the lack of immediately available newborn screening strategies.[4] Along these lines, it has been reported that the average time to perform the Kasai portoenterostomy in the United States varies between 61 and 63 days,[2,4,6] whereas in Europe it ranges between 57 and 68 days.[4] In this respect, the use of stool color charts in newborns with jaundice is a simple, non-invasive and low-cost screening strategy that has shown good results in several countries and could contribute to reduce the time to diagnosis of BA and, therefore, improve the prognosis of these children by allowing the performance of the Kasai procedure in the optimal time window for treatment.[1,7,11]

Notwithstanding the above, it is worth mentioning that in our study it was observed that the report of BA cases in which the age of the patient at the time of notification was less than 1 year progressively increased between 2018 and 2021 (28% vs. 55%). However, there is still room for making progress in the detection times of this condition, so it is

necessary that the relevant authorities in the country target their efforts to improve screening strategies and the level of knowledge of health personnel on this issue to speed up diagnosis times and, therefore, referral to bypass surgery.

The main limitation of the present study is the secondary nature of the data analyzed (information provided by the INS from the notification forms of BA cases in the country), as this made it impossible to access relevant information such as the exact age at diagnosis (the source only reports the age at the time of notification of the case to the INS, which was obtained from the date of birth of the patient and the date of notification of the case) or detailed clinical data of the patients both at the time of diagnosis and notification. Nevertheless, this is the first study that provides data on the incidence of this condition in the country, and it may be used as a starting point for further studies on the subject.

Conclusions

The incidence of BA in Colombia during 2018-2021, based on the number of cases in which the age of official notification was less than 1 year, is lower than what has been reported in other countries and regions of the world. In addition, the departments of Boyacá, Santander, and Cauca had the highest incidence rates.

Given the characteristics of the database analyzed, it is not possible to establish the age at diagnosis, but it is noteworthy that the median age at the time of notification of the event (4.89 months) is much higher than the optimal time window (first 60 days of life) for the initiation of the gold standard treatment (Kasai procedure), which implies a worse prognosis for these children. In this sense, it is necessary to improve screening strategies for this condition in newborns and to develop and implement health education interventions aimed at improving early diagnosis rates, correct notification, and timely care for these patients.

Conflicts of interest

None stated by the authors.

Funding

None stated by the authors.

Acknowledgments

To the Public Health Surveillance and Control of Non-Communicable Diseases Group of the Instituto Nacional de Salud, for making available the database for the analysis and preparation of the study.

References

1. Godoy M, López C, Álvarez-Chavez F, Borges-Pinto R, Botero-Osorio V, Dolz-Aguilar MV, *et al.* Colestasis neonatal: revisión narrativa del grupo de trabajo de la Sociedad Latinoamericana de Gastroenterología, Hepatología y Nutrición Pediátrica. Acta Gastroenterol Latinoam. 2022;52(3):344-54. https://doi.org/nzp8.
2. Feldman AG, Sokol RJ. Neonatal Cholestasis: Updates on Diagnostics, Therapeutics, and Prevention. Neoreviews. 2021;22(12):e819-36. https://doi.org/nzp9.
3. Siddiqui AI, Ahmad. T. Biliary Atresia. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2024 [cited 2024 Jan 7]. Available from: https://www.ncbi.nlm.nih.gov/books/NBK537262/.

4. Fawaz R, Baumann U, Ekong U, Fischler B, Hadzic N, Mack CL, *et al.* Guideline for the evaluation of cholestatic jaundice in infants: Joint recommendations of the North American society for pediatric gastroenterology, hepatology, and nutrition and the European society for pediatric gastroenterology, hepatology, and nutrition. J Pediatr Gastroenterol Nutr. 2017;64(1):154-68. https://doi.org/f3vj47.
5. Jimenez-Rivera C, Jolin-Dahel KS, Fortinsky KJ, Gozdyra P, Benchimol EI. International Incidence and Outcomes of Biliary Atresia. J Pediatr Gastroenterol Nutr. 2013;56(4):344-54. https://doi.org/f4s778.
6. Hopkins PC, Yazigi N, Nylund CM. Incidence of Biliary Atresia and Timing of Hepatoportoenterostomy in the United States. J Pediatr. 2017;187:253-7. https://doi.org/nzqb.
7. Chung PHY, Zheng S, Tam PKH. Biliary atresia: East versus west. Semin Pediatr Surg. 2020;29(4):150950. https://doi.org/nzqc.
8. Colombia. Congreso de la República. Decreto 1954 de 2012 (septiembre 19): Por el cual se dictan disposiciones para implementar el sistema de información de pacientes con enfermedades huérfanas [Internet]. Bogotá D.C.: Diario Oficial 48558; september 19 2012 [cited 2025 Jan10]. Available from: https://bit.ly/3DQIpY5.
9. Colombia. Departamento Administrativo Nacional de Estadística (DANE). Datos de nacimientos en Colombia [Internet]. Bogotá D.C.: DANE; 2024 [cited 2025 Jan 7]. Available from: https://bit.ly/4akBAdz.
10. Yerina SE, Ekong UD. Biliary Atresia/Neonatal Cholestasis: What is in the Horizon? Pediatr Clin North Am. 2021;68(6):1333-41. https://doi.org/gqd8cr.
11. Suarez-Galvis MM, Villa-Gómez CC, Santamaria-Escobar A, Ruiz-Navas P, Zuleta-Tobón JJ. ¿Reconocemos adecuadamente la acolia/hipocolia en los lactantes? Salud, Barranquilla. 2022;38(3):774-85. https://doi.org/nzqd.