v. 4, n. 70, 2024

International Journal of Health Science

DUODENAL ATRESIA ASSOCIATED WITH ANNULAR PANCREAS IN A PATIENT WITH DOWN SYNDROME CASE REPORT IN A PUBLIC HOSPITAL IN THE STATE OF GOIÁS

Júlia Pires de Farias
``*Universidade Federal de Jataí* `` (UFJ)
Jataí - GO
https://orcid.org/0000-0002-1176-6349

Arícia Mota
``*Universidade Federal de Jataí* `` (UFJ)
Jataí - GO
https://orcid.org/0000-0001-6254-583X

André Gebrim Matias
``*Universidade Federal de Jataí* `` (UFJ)
Jataí - GO
https://orcid.org/0000-0002-3351-1005

Sara Alves França
``*Universidade Federal de Jataí* `` *(*UFJ)
Jataí - GO
https://orcid.org/0000-0002-8382-1394

Gabriel Ramos Macedo Moraes
``*Universidade Federal de Jataí* `` ``(UFJ)
Jataí - GO
https://orcid.org/0009-0000-9434-9315

Pedro Ricardo Mendes Llobet
``*Universidade Federal de Jataí* `` ``(UFJ)
Jataí - GO
https://orcid.org/0000-0002-1090-2820

Mathias Rezende Macedo
``*Universidade Federal de Jataí* `` (UFJ)
Jataí – GO
https://orcid.org/0000-0001-5427-3344

Monique Costa Dias
``*Universidade Federal de Jataí* `` *(*UFJ)
Jataí - GO
https://orcid.org/0000-0002-9521-062X

Sales José Lopes Gonçalves Rosa
``*Universidade Federal de Jataí* `` (UFJ)
Jataí - GO
https://orcid.org/0000-0001-6120-4611

Denise da Costa Carvalho
``*Universidade Federal de Jataí* ``(UFJ)
Jataí - GO
http://lattes.cnpq.br/2041296581865966

Abstract: Duodenal atresia is a congenital disorder that corresponds to the most common cause of intestinal obstruction in newborns. This relationship is even more present in patients with trisomy 21, who can also suffer from other malformations related to the gastrointestinal tract, such as annular pancreas. Infants who present with duodenal atresia commonly present with abdominal distension, dietary intolerance and vomiting in the first hours of life. The suspected diagnosis can be initiated prenatally with ultrasounds (USG), but confirmation usually occurs in the postnatal period through visualization of the "double bubble" sign on a simple abdominal x-ray. The treatment of this condition is surgical, with duodenoduodenostomy usually being performed to divert intestinal transit and correct the obstruction. This is a case report of duodenal atresia associated with annular pancreas in a patient diagnosed with Down syndrome. The case discussion was based on 12 articles selected from the Pubmed, Scielo, UpToDate and VHL databases. The study is descriptive and analytical, exploring the characteristics observed in the case report and related to the literature description. The report is of a male neonate, born by cesarean section with 35 weeks and 4 days of gestational age due to Doppler flowmetry suggesting zero diastole. During prenatal care, a morphological USG had already been performed, showing a double bubble sign, which is why an oroenteral tube was introduced in the delivery room, with good progression. The newborn (NB) presented typical facial features, with an oblique palpebral fissure, flat face and depressed nasal bridge. On the second day of life he developed bilious vomiting. Imaging tests showed a double bubble sign and genetic evaluation revealed a karyotype of 47 The patient evolved with good acceptance of the oral diet and was discharged after 37 days of life with good weight gain.

Keywords: Congenital Malformations; Trisomy; Pancreatopathies.

INTRODUCTION

Duodenal atresia is a congenital intestinal obstruction and is one of the most common diagnoses of gastrointestinal obstruction diagnosed in newborns, occurring in 2 in every 10,000 live births. Its exact etiology is still unknown, although there are theories that believe that failure to recanalize the solid epithelial cord or excessive endodermal proliferation are the pathophysiological explanation for duodenal atresia (SIGMON, D. et al., 2024).

It is constantly associated with other anomalies, such as Down syndrome, and around 30 to 40% of newborns affected by duodenal atresia have trisomy 21. Furthermore, there are also relationships with annular pancreas and atresia of other intestinal portions, such as jejunum, ileum and rectum (SIGMON, D. et al., 2024).

The annular pancreas is a congenital malformation that is characterized by the expansion of a portion of pancreatic tissue involving part of the duodenum, causing extrinsic duodenal obstruction. The symptoms produced by this comorbidity are similar to those of duodenal atresia, often indistinguishable, making it difficult to diagnose the two conditions when they are associated (GONÇALVES, M. et al., 2008).

Diagnostic suspicion can be raised both prenatally, with ultrasound evidence of the double bubble sign, and postnatally, with the presence of clinical signs and symptoms of upper intestinal obstruction, with bilious vomiting being more evident, generally in the first hours. of life (LOPEZ-DIAZ, N. et al., 2023). However, diagnostic confirmation can only be achieved postnatally using simple abdominal radiography, barium fluoroscopy and computed tomography (SIGMON, D. et al., 2024).

The management of duodenal atresia is surgical, and bypass of the affected segment through a duodenoduodenostomy is generally indicated in neonates to relieve intestinal obstruction.

In cases associated with an annular pancreas, the aim of surgery is to alleviate the outflow duodenum bypassing the pancreatic ring, since its resection must be avoided, as it is often associated with serious complications such as pancreatitis and fistula formation (THUKRAL, C. et al., 2024).

As the association of duodenal atresia and annular pancreas is uncommon, the description of this clinical case is justified as a contribution to better elucidating these conditions.

CLINICAL CASE

IDENTIFICATION

Neonate, male, late preterm, born by cesarean section of 35 weeks and 4 days (First trimester ultrasound (USG) at 7 weeks + 3 days), with indication of obstetric delivery via discharge by Doppler flowmetry suggesting zero diastole. The Apgar was 8 in the first minute and 10 in the fifth minute. Anthropometric data indicated a weight of 2,250g, length of 44 centimeters, 32 cm head circumference, 29 cm chest circumference, 27 cm abdominal circumference, therefore being suitable for gestational age (AGA). Umbilical cord clamping was performed early (<1 minute), followed by the need for airway aspiration and external stimulation.

GESTATIONAL DATA

Maternal age 32 years, single, planned pregnancy, negative serology and immunity to toxoplasmosis, rubella and cytomegalovirus. Routine first trimester exams were carried out, which revealed gestational hypothyroidism, treated with Levothyroxine 50 mcg until the

date of delivery. The pregnant woman took folic acid and ferrous sulfate supplementation. During the second trimester, a morphological USG was performed, at 23 weeks and 5 days, in which a double gastric bubble sign was identified, which could correspond to a case of duodenal atresia.

EVOLUTION

In the delivery room, an oroenteral tube was inserted into the newborn, with good progression, remaining under observation in the post-anesthesia recovery room with the parturient.

Mother-neonate binomial were referred to rooming-in, where they remained in the care of the hospital team. The diet for newborns (NB) was released with guidance for the nursing team and parents regarding monitoring gastrointestinal symptoms. On the second day of life, the newborn began to experience nausea and vomiting, "black-brown", and was taken to the Neonatal Intermediate Care Unit (NICU). He was eupneic on room air, with a distended and hypertympanic abdomen, zona 3 of Krammer, typical facies, with oblique palpebral fissure, flat face and depressed nasal bridge. He was using an orogastric tube with a total bilious/brown output of 56 mL/day.

SUBSIDIARY EXAMS

In imaging studies, REED (esophageal, stomach and duodenum radiography) showed: gastric distension and an image suggestive of a double bubble, and in abdominal USG: mild ectasia of the intrahepatic bile ducts in the left hepatic lobe and hyperdistended gallbladder. Laboratory tests indicated total bilirubin 11.9 and indirect bilirubin 11.5, in addition to genetic evaluation that revealed the karyotype 47XY +21, compatible with Down syndrome.

Transfontanelle USG showed no changes. Cardiological auscultation revealed a systolic murmur on the left sternal border, submitted to a Transthoracic Echocardiogram which identified a wide interventricular communication with the aorta overriding the septum and hypertrophy of the right ventricle, suggesting tetralogy of Fallot with good anatomy (Pink Fallot).

CONDUCT

Under the care of the neonatal intensive care unit (NICU) team, he remained hemodynamically stable, with good oxygen saturation (94-95%) on room air and zero enteral nutrition. She received pediatric surgery evaluation with surgical programming, underwent antibiotic prophylaxis, phototherapy and parenteral nutritional support. Furthermore, it was evaluated by cardiology, which provided surveillance guidelines regarding saturation during hospitalization and surgical procedures and the possibility of monitoring the pathology on an outpatient basis, given the good anatomy of the change.

After stabilization of the clinical condition, an exploratory laparotomy was performed in the eighth day of life, which identified duodenal obstruction due to annular pancreas at the level of the 2nd portion of the duodenum, proceeding with lysis of congenital abutments and duodenoduodenum anastomosis to correct intestinal narrowing. In the NICU, he remained on zero enteral diet until the 4th postoperative day (PO), when an enteral diet via nasogastric tube was started with good acceptance. On the 17th day of PO, he was taken to the NICU still using a nasogastric tube for diet and guidance on breastfeeding stimulation. Given 29 days PO and 37 days of life, the newborn is discharged with an oral diet, good weight gain and good surgical wound healing, using furosemide 2.09 mg/kg/day, spironolactone 1.12 mg/kg /day, multivitamin 12 drops/day and zinc 1.12 mg/kg/day, remaining hemodynamically stable,

without gastrointestinal symptoms and with guidelines for follow-up and outpatient clinical care.

DISCUSSION

Duodenal atresia is a complete intestinal obstruction that is the most common cause of congenital gastrointestinal obstruction diagnosed in neonates. Its occurrence is approximately 1:10,000 live births, accounting for around 60% of cases of intestinal atresia (KING, A., 2024). Its etiology is not completely elucidated, but it is understood that atresia occurs due to failure to restore the permeability of the intestinal tract during the period from the 8th to 10th weeks of embryogenesis, due to excessive proliferation of the endoderm or failure to recanalize the solid epithelial cord. (SIGMON, DF; EOVALDI, BJ; COHEN, HL, 2024).

It is subdivided into three types: type 1 (69% of cases) – mucous membrane atresia, without muscle involvement; type 2 – fibrous cord connecting the duodenum in two portions; type 3 – complete separation of the duodenum halves with abnormalities of the biliary tract (BULAS, DI, 2024).

Some risk factors are associated with its occurrence, such as cystic fibrosis, gastroschisis, midgut volvulus, use of maternal medications (such as vasoconstrictors), hereditary thrombophilias and other genetic changes.

Trisomy 21 (Down syndrome) is the congenital anomaly most frequently associated with cases of duodenal atresia. It is estimated that 28 to 40% of cases of duodenal atresia have Down syndrome, a situation found in the reported case. (SIGMON, DF; EOVALDI, BJ; COHEN, HL, 2024). It is noteworthy that, among children with Down syndrome, 5% of them are at risk of gastrointestinal tract anomalies, with duodenal atresia being the most common, corresponding to around 2.5% of cases (OSTERMAIER, K., 2024).

In some cases, such as the neonate in this report, duodenal atresia may be associated with an annular pancreas, a rare congenital alteration that consists of the presence of a ring of pancreatic tissue surrounding the descending portion of the duodenum, resulting from incomplete rotation of the ventral band of the pancreas. around the duodenum during embryogenesis, around the 7th week. It is classified as complete and incomplete, depending on the degree of involvement of the 2nd duodenal portion, and also in grades I to VI, depending on the site of drainage of the annular duct, with types I and II being more common, draining directly into the pancreatic duct. main or to the major papilla (THUKRAL, C.; FREEDMAN, SD, 2024).

Even during prenatal care, gastrointestinal anomalies can be diagnosed and care planned after birth. Through USG it is possible to visualize the "double bubble" sign, which corresponds to the stomach without echo, full of amniotic fluid (first bubble), and a second, more distal nearby structure, filled with fluid, often circular (but with a blunt end); which is the obstructed portion of the duodenum - the second bubble. This ultrasound finding can occur when, in the presence of obstruction in the proximal small intestine, more frequently in duodenal atresia, there is dilation of the stomach and duodenal bulb (SIGMON, DF; EOVALDI, BJ; COHEN, HL, 2024).

In a cohort study carried out to evaluate gastrointestinal anomalies, a total of 18 monitored pregnancies, which showed a double bubble sign during prenatal care, obtained postnatal confirmation of duodenal atresia during surgical exploration and corrective procedures. This suggests that the double bubble sign is a reliable positive predictor of duodenal atresia and other etiologies are probably rarer (BISHOP, Juliet C. et al., 2020).

The suspicion of congenital intestinal atresia can be clinically raised after birth, due to the occurrence of vomiting in the first 24 to 38 hours of life, especially after the first feeding, with progressive worsening if untreated. Vomiting can be bilious or non-bilious, the latter being related to an obstruction proximal to the greater duodenal ampulla and the former to obstruction distal to the ampulla (SIGMON, DF; EOVALDI, BJ; COHEN, HL, 2024). In general, the symptoms resemble classic intestinal obstruction, progressing with vomiting, food intolerance and progressive abdominal distension (KING, A.). The case reported here also presents an annular pancreas, which corroborates the clinical signs of obstruction, occurring in 1/3 of patients with the condition (THUKRAL, C.; FREEDMAN, SD, 2024).

In the complementary evaluation, the test that allows the earliest diagnosis of duodenal atresia, as previously mentioned, is prenatal USG, with the finding of a "double gastric bubble". However, this ultrasound evidence occurs in less than half of the cases (KING, A.), making postnatal investigation necessary. After birth, simple abdominal radiography, barium fluoroscopy – which shows intestinal obstruction– and, more restrictedly, computed tomography can be used (SIGMON, DF; EOVALDI, BJ; COHEN, HL, 2024). The same finding as double bubble can be seen on abdominal radiography (GONÇALVES, MEA; VIANA, DC; RODRIGUES, JC; MACEDO, DB; LIMA, TL, 2008). A tomography is rarely used, since its challenging technique in neonates requires sedation and intravenous administration of contrast, and is reserved for inconclusive cases of intestinal obstruction (SIGMON, DF; EOVALDI, BJ; COHEN, HL, 2024).

Definitive treatment consists of surgical correction of the condition, and between birth and the procedure, treatment is only supportive, performing aspiration with a nasogastric tube for stomach decompression (SIGMON, DF; EOVALDI, BJ; COHEN, HL, 2024). The surgery consists of a bypass of the compromised segment through a duodenoduodenostomy or a duodenojejunostomy (VINYCOMB, Toby et al., 2020). The procedure can be performed open or laparoscopically (SIGMON, DF; EOVALDI, BJ; COHEN, HL, 2024).

If available, the minimally invasive surgeries reduce postoperative symptoms and conditions, such as intestinal adhesions, pain and length of hospital stay, in addition to better aesthetic result (GUELFAND CH, MIGUEL et al., 2024). In the case in question, through exploratory laparotomy it was possible to confirm the duodenal atresia evidenced prenatally, as well as identify the presence of the annular pancreas at the level of the 2nd portion of the duodenum.

Postoperatively, patients require continuous nasogastric drainage, which may or may not be accompanied by parenteral nutrition. Once the output from the nasogastric tube significantly decreases or stops, feeding can be started in low quantities and advanced as tolerated, with no contraindication to breastfeeding and stimulation of sucking being indicated (SIGMON, DF; EOVALDI, BJ; COHEN, HL, 2024).

The possible complications of surgical correction are maximum in the immediate postoperative period, with a reduction in occurrence in the long term, including: duodenal motility disorders, adhesive intestinal obstruction and gastroesophageal reflux disease (VINYCOMB, Toby et al., 2020). After surgical treatment, the patient must be monitored on an outpatient basis. The prognosis after resolving surgical treatment of duodenal atresia is excellent, with evaluated studies demonstrating late complications in up to 12% of patients and late mortality of 6% (SIGMON, DF; EOVALDI, BJ; COHEN, HL, 2024).

Added to this, there were no fetal or neonatal deaths in cases of isolated duodenal atresia, which supports the fact that fetuses with isolated duodenal atresia on prenatal USG show favorable results. However, cases with additional anomalies have less favorable outcomes, since the only fetal or neonatal deaths observed occurred in cases of duodenal atresia with Down syndrome and associated additional structural anomalies (BISHOP, Juliet C. et al., 2020).

In these cases, the main causes of morbidity and mortality are prematurity, low birth weight and associated anomalies, such as short bowel syndrome and cardiac anomalies, common conditions in children with Down syndrome (VINYCOMBA, T., 2019). The prognosis of complex atresias depends on the length and function of the remaining intestine (KING, A., 2024).

Regarding the impact of this pathology on patients' quality of life, it was shown that children undergoing surgical correction of duodenal atresia early in life normally present quality of life results comparable to healthy ones. However, children with trisomy 21 associated with duodenal atresia have long-term needs that differ from other patients with duodenal atresia, due to the high association of this trisomy with other congenital anomalies (VINYCOMBA, T., 2019).

Regarding the annular pancreas, there are few detailed and comparative studies, due to the rarity of the condition. However, the results found in the literature show that duodenal diversion and pancreatic resection are effective and safe alternatives in the long term, when well indicated (SCHMIDT, Marcelo K. et al., 2004).

CONCLUSION

Thus, the work proposed to describe a preterm neonate, diagnosed with Down syndrome, in which two malformations of the gastrointestinal tract occurred that lead to intestinal obstruction and are associated with trisomy 21. It is concluded that it is important to investigate intestinal atresia in all newborns who develop obstructive symptoms of the gastrointestinal tract in the first days of birth, even in those in whom prenatal USG did not identify changes. The importance of a surgical approach to correct the alteration and early return to enteral nutrition is highlighted, thus avoiding complications related to the use of prolonged parenteral nutrition. Furthermore, it is important to do more work in the literature on the annular pancreas, such as case reports, describing diagnosis, surgical approach and evolution, as well as longitudinal work monitoring these patients.

REFERENCES

BISHOP, Juliet C. et al. The double bubble sign: duodenal atresia and associated genetic etiologies. **Fetal diagnosis and therapy**, v. 47, n. 2, p. 98-103, 2020.

DU PREEZ, H.; BRITS, E. **The profile and outcome of small bowel atresia at Universitas Academic Hospital**. South African Journal of Surgery, v. 61, n. 1, p. 31-35, 2023.

SIGMON DF, EOVALDI BJ, COHEN HL. Duodenal Atresia and Stenosis. [Updated 2023 Jun 26]. **StatPearls [Internet]: StatPearls Publishing**; 2024 Jan.

GUELFAND CH, MIGUEL et al. Cirugía mínimamente invasiva en recién nacidos de< 2.500 g. **Revista chilena de pediatría**, v. 85, n. 1, p. 64-67, 2014.

GONÇALVES, Manoel Eduardo Amoras et al. Atresia duodenal associada a pâncreas anular: relato de caso. Rev. para. med, 2008.

LÓPEZ-DÍAZ, Néstor Gibrán; OLIVER-GARCÍA, Edgar Fernando; NÚÑEZ-ENRÍQUEZ, Juan Carlos. Manejo quirúrgico de atresia duodenal por páncreas anular y atresia intestinal IIIb. **Revista Médica del Instituto Mexicano del Seguro Social**, v. 61, n. 1, p. 123, 2023.

SCHMIDT, Marcelo Kruel et al. Pâncreas anular: ressecção pancreática ou derivação duodenal. **Revista da Associação Médica Brasileira**, v. 50, p. 74-78, 2004.

VINYCOMB, Toby et al. Quality of life outcomes in children born with duodenal atresia. **Journal of Pediatric Surgery**, v. 55, n. 10, p. 2111-2114, 2020.

C Thukral, MD, PhD, & S D Freedman, MD, PhD. (2024). Annular pancreas. In D G Adler, & S Grover (Ed). **UpToDate**. Acessado em 12 de maio.

A King, MD. (2024). Intestinal atresia. In J A Garcia-Prats, MD, M B Heyman, MD, & A G Hoppin, MD (Ed). **UpToDate**. Acessado em 12 de maio.

K K Ostermaier, MD. (2024). Down syndrome: Clinical features and diagnosis. In J E Drutz, MD, H V Firth, DM, & N Tehrani, MD (Ed). **UpToDate**. Acessado em 12 de maio.

D I Bulas, MD. (2024). Prenatal diagnosis of esophageal, gastrointestinal, and anorectal atresia. In D Levine, MD, L Wilkins-Haug, MD, & V A Barss, MD. **UpToDate**. Acessado em 12 de maio.