
PROVEN CASES OF DUODENAL ATRESIA ON PLAIN ABDOMINAL RADIOGRAPHY IN CORRELATION WITH SURGICAL FINDINGS – A CASE SERIES

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ABSTRACT

Duodenal atresia is a congenital emergency abnormality and the most common cause of proximal bowel obstruction in neonates. The duodenum has not developed properly in embryogenesis with a blind end that cannot be seen through gastric contents. The incidence of duodenal atresia is 1/10.000 live births that commonly affects boys than girls. In cases of neonatal bilious vomiting, the first choice imaging modality is plain abdominal radiography. The purpose of this case report is to increase radiologist and pediatrician awareness in duodenal atresia diagnosis through plain abdominal radiography and to minimize radiation in the neonates. We report 3 cases of duodenal atresia found in Sanglah Hospital, Denpasar, in 1 year for the period 2018 - 2019. Case 1: A 4-day-old baby girl presented with bilious vomiting in the first 24 hours after birth. Case 2: A 4-day-old baby boy presented with lethargy, weak crying, and groaning after birth. There is no history of passed meconium in the first 24 hours of birth. Case 3: A newborn boy presented with bilious emesis after breastfeeding and nonprojectile. From the 3 cases, a plain abdominal radiograph was examined, and a *double bubble sign* with no intestinal gas distal was found.

Keywords: Duodenal Atresia; Bilious Vomiting; *Double Bubble*; Plain Abdominal Radiography; Duodenoduodenostomy.

INTRODUCTION

Duodenal atresia is a proximal congenital gastrointestinal obstruction that can cause bilious or non-bilious vomiting within the first 24-38 hours of birth, usually after the first oral feeding. There is interference during the embryogenesis process. It produces a short fibrous cord that connects between the two blind ends of the atretic duodenum. This is considered the result of duodenal failure for recanalization, which begins at the 6th week of embryogenesis development.¹ Duodenal atresia is a congenital abnormality that occurs in about 1/10.000 live births, which commonly affects boys than girls. More than 50% of duodenal atresia is associated with

another congenital anomaly. Duodenal atresia is located at the junction of the 1st and 2nd parts of the duodenum, found in 85% of cases.² Classification of duodenal atresia based on Gray and Scandalakis is divided into three types. Type I is the most common type and often occurs in neonates. Type II is the rarest type, about 1%, and type III occurs in about 7% of cases.

The diagnosis of duodenal atresia can be made radiographically with a plain abdominal radiograph as the first step in evaluation. The double bubble sign with the absence of distal intestinal gas is a pathognomonic sign for duodenal atresia.³

CASE SERIES

Case 1

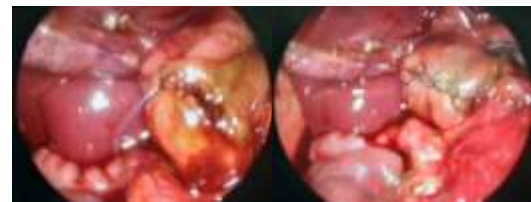
A 4-day-old baby girl presented with bilious vomiting within the first 24 hours of birth, typically following after breastfeeding. There was no history of passed meconium within the first 24 hours after birth. The patient had a history of cesarean delivery et causa re-cesarean section delivery with normal gestational age, the 4th child, and birth weight is 3.100 gram and normal APGAR score.

On physical abdominal examination, there are supel, no distention, and normal bowel sound. 20 cc residual from the orogastric tube was obtained. There is a normal laboratory blood examination. Plain abdominal radiography showed a double bubble sign in the middle and left abdominal cavity with the absence of distal bowel gas, so impressed as duodenal atresia.

Duodenoduodenostomy laparoscopic is performed on the next day, duodenal atresia in surgical findings, and anastomosis end to end is done.



a. Plain abdominal radiography showed a double bubble sign in the middle and left abdominal cavity and no distal intestinal gas.



b. Duodenal atresia finding on duodenoduodenostomy laparoscopic and anastomosis end to end is done.

Figure 1. Plain abdominal radiography and surgical findings in case 1.

Case 2

A 4-day-old baby boy presented with lethargy, weak crying, and groaning after birth. APGAR score 5-8-8. Respiratory distress (-), cyanosis (-). 13 cc bilious material production in an orogastric tube after birth was obtained. No history of passed meconium within the first 24 hours of birth. The patient is the 2nd child, with premature gestational age (31 - 32 weeks) and spontaneous delivery history, birth weight is 1.550 gram.

On physical abdominal examination, there are supel, no distention, and normal bowel sound. Bilious material production in the orogastric tube remained. On plain abdominal radiography showed double bubble sign with the absence of bowel gas distally, so impressed as duodenal atresia.

Surgical was performed and founded duodenal atresia. Duodenoduodenostomy with Kimura procedure is done.



Figure 2. The plain abdominal radiograph showed a double bubble sign with the absence of bowel gas distally.

Case 3

A newborn boy presented with bilious emesis after breastfeeding and nonprojectile. No history of passed meconium within 24 hours after birth. The patient is the 1st child with cesarean delivery et causa polyhydramnios, birth weight is 2.680 gram, and a normal APGAR score. Antenatal ultrasonography was obtained and revealed a double bubble appearance.

On physical abdominal examination, there are supel, no distention, and normal bowel sound. On the plain abdominal radiography showed double bubble sign with the absence of bowel gas distally, so impressed as duodenal atresia.

Atresia 2nd part of the duodenum was founded in duodenoduodenostomy surgical with Kimura procedure and end to end anastomosis duodenoduodenostomy diamond shape is done.



a. Plain abdominal radiography showed a double bubble sign and no distal bowel gas.



b. Atresia 2nd part of duodenum finding on duodenoduodenostomy surgical.

Figure 3. Plain abdominal radiography and surgical findings in case 3.

DISCUSSION

In newborns with vomiting, especially bilious vomiting, should be considered the

possibility of proximal bowel obstruction. Other clinical symptoms and investigation examinations support it are carried out to establish the diagnosis. In this case, the imaging modality plays an important role in establishing the diagnosis. Complete history accompanied by information about the antenatal examination in duodenal atresia can also be an adjunct in establishing diagnoses such as polyhydramnios and double bubble sign on antenatal ultrasound. The three serial cases of duodenal atresia were obtained in the past year that was found at Sanglah Hospital, with bilious vomiting and no meconium passed in the first 24 hours of birth.

The duodenum is a small intestine C-shaped that connects the stomach to the jejunum. The duodenum arches around the head of the pancreas and forms the terminal or orifice of the liver and pancreas' biliary apparatus system. Besides, the duodenum is also the end of the upper gastrointestinal tract. Gastrointestinal is divided into upper and lower parts by the presence of Treitz ligament (*M. suspensorius duodeni*), which is located in the duodenojejunalis flexura, which is the boundary between the duodenum and jejunum. The duodenum is located in the abdominal cavity in the epigastric and umbilical regions. The duodenum is divided into four parts: superior part, descendens part, horizontalis part, and ascendens part.⁴ Development of duodenal embryology begins at the 5th week of pregnancy, where the proliferating duodenal epithelium will completely block the duodenal lumen in the 6th week. At the 7th week there will be a vacuolization process. During this process, the cell will undergo an apoptosis process that arises in the lumen of the duodenum. Apoptosis will cause epithelial cell degeneration. This process results in recanalization of the duodenal lumen in the 9th week.

Atresia is a medical term describing a condition where no opening or part of the body is closed.⁵ Duodenal atresia is defined as a condition in which the duodenum does not develop properly, so it is not an open channel of the stomach that does not allow food to travel from the stomach to the intestine. In this

condition, the duodenum can experience complete narrowing, inhibiting food absorption from the stomach into the intestine.⁶

Congenital abnormalities are one of the main causes of infant death. Based on WHO, more than 8 million babies worldwide are born every year with congenital abnormalities WHO states that congenital abnormalities cause 2.68 million infant deaths, 11,3%. When compared with Southeast Asia, Indonesia is still the country with a high prevalence of babies with congenital abnormalities. The Ministry of Health's surveillance results for the period of September 2014 - March 2018 showed 1.085 infants with reported congenital abnormalities. However, there was no specific data regarding intestinal atresia epidemiology because it was still not well recorded.⁷ Congenital duodenal atresia occurs in about 1 / 10.000 live births and affecting boys more often than girls.

There are two suspected factors causing duodenal atresia, namely, intrinsic and extrinsic factors. Intrinsic factors are caused due to failure of epithelial nerve recanalization or excessive endodermal proliferation.⁸ In contrast, the extrinsic factor of duodenal atresia is caused by the development of the disturbance of the surrounding organ structure, such as the pancreas. The annular pancreas is a pancreatic tissue surrounding the duodenum, especially the duodenum descendens part.⁹

More than 50% of duodenal atresia is associated with congenital anomalies, namely trisomy 21 / Down Syndrome (about 30% of patients), as part of the VACTERL anomaly complex (Vertebral defects, Anal atresia, Cardiac defects, Tracheal Esophageal fistula, Renal anomalies, and Limb abnormalities); 30% isolated heart defects; 45% prematurity; 33% growth retardation; 25% other intestinal anomalies.²

The classification of duodenal atresia, according to Gray and Scandalakis is divided into three types, including :¹⁰

Type I, characterized by the presence of a web or membrane that obstructs the duodenal lumen. There are three types of membrane abnormalities

in this type, namely simple, fenestration and windsock anomalies. Windsock anomalies may occur if the web is thin. The base of this membrane is the 2nd part of the duodenum. This type is the most common type of all duodenal atresia (about 92%). This type of atresia may partially obstruct and can, therefore, not be detected until solid food is given. **Type II**, the two blind ends of the duodenum with the proximal and distal segments connected by short fibrous cords. This type is the least common type of duodenal atresia, about 1%. **Type III**, the two blind ends of the duodenum with the complete discontinuity between the proximal and distal segments, occurs in 7% of duodenal atresia cases.



Gambar 4. Classification of duodenal atresia according to Gray and Scandalakis. (A) Type I duodenal atresia; (B) Type II duodenal atresia, and (C) Type III duodenal atresia.²

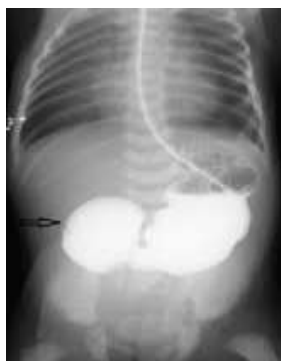
Duodenal atresia appears early in the birth with bilious vomiting, usually within the first 24-38 hours of birth after the first drink, and will worsen if untreated. Infants with duodenal atresia also appear in the early period with minimal abdominal distension, and 60-75% of neonates have no meconium release within the first 24 hours of birth. If there is a meconium release in neonates with atresia, usually a small amount of meconium, a drier consistency, and a grayish color than normal meconium. Obstruction often occurs around the ampulla of Vater. If atresia occurs in the distal ampulla of Vater in the 2nd part of the duodenum, bilious vomiting occurs. If atresia occurs proximal to the ampulla of Vater, non-bilious vomiting occurs. Dehydration, weight loss, and electrolyte imbalance can occur immediately unless fluid and electrolyte loss is replaced adequately.¹¹ If the condition is not

treated quickly, hypocalcemia metabolic alkalosis or hypochloremic can occur.

Plain abdominal radiography revealed a double bubble sign accompanied by the absence of gas in the distal part intestinal, depicting obstruction of the stomach and duodenum, which is pathognomonic for duodenal diagnosis atresia.¹² If the stomach or duodenum is decompressed by an orogastric tube or vomiting occurs, a double bubble sign may not be seen on the initial radiography.



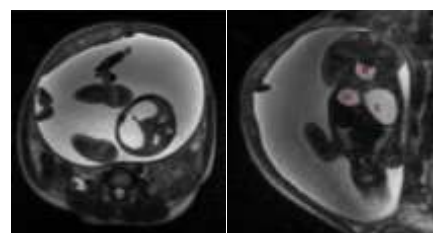
a. Plain "double bubble sign" abdominal radiographs of duodenal atresia.



b. Upper gastrointestinal contrast study with a double bubble sign and no contrast seen distally.¹³



c. Double bubble sign of duodenal atresia on abdominal ultrasound (ST = stomach; D = duodenum) (13).



d. T2-weighted axial (left) and coronal (right) images of foetus showing distention stomach and duodenal bulb giving a double bubble sign accompanied by an increased amniotic fluid suggestive of polyhydramnios (S = stomach; D = duodenal bulb; B = bladder).¹⁴

Gambar 5. Plain abdominal radiography, contrast examination, abdominal ultrasound, and abdominal MRI in duodenal atresia

Plain abdominal radiography is and continues to be a good and useful tool in diagnosing neonatal intestinal obstruction. The sensitivity of plain abdominal radiography in diagnosing neonatal gastrointestinal obstruction is 85,2%. In Malhotra's study in 1997, plain radiography was diagnostic in 50-60% of neonatal small bowel obstruction, equivocal in 20-30%, and non-diagnostic in 10-20%.¹⁵ In neonates with a classic double bubble sign, an additional radiological examination is not required. The surgeon is prepared to plan the surgery because all congenital cause of duodenal obstruction requires surgery.¹⁶ An upper gastrointestinal contrast study shows contrast filling of distended gastric and proximal duodenum with no visible contrast distally. However, this examination is usually not performed because plain radiography is basically diagnostic. Duodenal atresia on abdominal ultrasound is also seen as a typical double bubble sign, depicting a fluid-filled stomach and proximal duodenum separated by narrowing at the location of the pylorus that is wider than normal.¹²

Duodenal atresia in neonates can be diagnosed antenatally. A serial antenatal examination can detect duodenal obstruction

in the 3rd trimester of pregnancy. Polyhydramnios occurs during the 3rd trimester in almost all cases of duodenal atresia. So the diagnosis of antenatal duodenal atresia can be made at 32-36 weeks gestation for cases with polyhydramnios.¹¹ On ultrasonography and magnetic resonance imaging (MRI), a typical double bubble sign is seen due to distention of the stomach and duodenal bulb. The remaining small intestine loops appear to collapse. Ultrasonography findings are enough to diagnose; however, MRI helps exclude several intestinal atresias that have different postnatal prognosis and management. T2-weighted MRI images are important in making a diagnosis that shows a double bubble sign associated with hyperintense fluid in the stomach and duodenal bulb at the level of obstruction. T1-weighted MRI images help to exclude the presence of additional atretic segments that shows meconium in the distal loop of the small intestine and colon.¹⁴

Before surgery, the stomach and proximal duodenum are decompressed using an orogastric tube, and intravenous fluid resuscitation is performed. Surgical correction by duodenoduodenostomy can be done with an open procedure or laparoscopy. The open approach is the most commonly used to correct duodenal atresia. The duodenum was mobilized using the Kocher maneuver. Duodenal atresia can also be corrected with duodenoduodenostomy as described by Kimura. Transversal duodenotomy is made in a dilated proximal segment and connected with a longitudinal duodenotomy along the distal portion to form a diamond shape.²

Recent developments in laparoscopic equipment and techniques have triggered changes in infant and pediatric surgical care. The advanced laparoscopic technique in neonates has recently led to a new surgical approach, laparoscopic duodenoduodenostomy. Comparison between laparoscopic and open operations procedure allows a reduction in intestinal function and

short transient ileus. Surgeons compared the laparoscopic and open approach on congenital duodenal obstruction that laparoscopic group can start feeding faster, full feeding, and shorter length of stay in hospital. The weakness of laparoscopic surgery that has been reported is postoperative leakage after conventional suturing techniques.¹⁷

Postoperative complications are uncommon, possible early complications include leakage of anastomosis and constriction,¹⁷ easy dehydration, functional duodenal obstruction, adhesion, and bowel movement problems.⁸ But advanced complications (megaduodenum, blind loop syndrome, GERD, esophagitis, pancreatitis, cholecystitis, and intraabdominal sepsis) occur in very rare cases.²

The prognosis of duodenal atresia with early surgical intervention is very good. With surgical treatment, neonates' survival rate with duodenal atresia reaches more than 90% (12). According to Milar (2005), although the prognosis of duodenal atresia is generally good, the mortality rate is 7%. High mortality is caused by prematurity and congenital abnormalities that accompany it.⁸

CONCLUSION

Duodenal atresia is a congenital abnormality that was included in neonates' emergencies and common in proximal neonatal gastrointestinal obstruction due to failure of recanalization during embryogenesis.¹⁰ This causes the stomach contents can't pass through so that the symptoms of vomiting arise, often bilious vomiting in the first 24 hours of birth accompanied by the absence of meconium.

Patients with duodenal atresia are often associated with other congenital abnormalities. Baby with duodenal atresia associated with down syndrome in 30% of cases. Early signs of duodenal atresia are minimal abdominal distension, bilious vomiting, and no meconium release in the first 24 hours of birth. The imaging modality has an important role in establishing the diagnosis. In this case, the plain abdominal radiography as an initial modality

that presents a double bubble sign and the absence of intestinal gas in the distal part, which is pathognomonic for duodenal atresia radiological examination is no need required. The sensitivity of plain abdominal radiography is 85,2% in diagnosing neonatal gastrointestinal obstruction. Selection of the right initial imaging can minimize radiation in neonates.

Continuous vomiting caused dehydration, weight loss, and electrolyte imbalance. Therefore adequate fluid and electrolyte replacement must be carried out. After the patient is stable, fluid and electrolyte replacement is resolved, surgery can be performed as a duodenal atresia treatment.

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