# Medical and Clinical Case Reports

# Late Diagnosis and Management of Type IV Jejunoileale Atresia in Low Income Setting

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Received: 05 Oct 2024; Accepted: 22 Nov 2024; Published: 03 Dec 2024

**Citation:** Kanyinda F, Talona R, Kasereka F, et al. Late Diagnosis and Management of Type IV Jejunoileale Atresia in Low Income Setting. Med Clin Case Rep. 2024; 4(4): 1-4.

# ABSTRACT

Intestinal atresia is a common cause of obstructive bowel in neonates. The jejuno-ileum is the most common site, followed by duodenal and colonic atresia. Late consultation in our setting has a negative impact on the prognosis. We report a case of a 7 days female newborn who was referred to the teaching hospital of University of Kisangani from a health center with symptoms related to bowel obstruction. Diagnosis and management were done at the department of surgery under limited resources.

#### **Keywords**

Diagnosis, Management, Jejunoileal atresia, Low income setting, DR Congo.

# Background

Intestinal atresia is a common cause of neonatal bowel obstruction (NBO). The incidence varies from 1/1,500 to 1/330 live births [1,2]. The most common site is the jejuno-ileum [3,4] and it results from ischaemic injury during pregnancy due to a mesenteric vascular accident leading to obliterative scarring [5,6].

There are four type of intestinal atresia: type I (mucosal), type II (atretic fibrous cord), type IIIa (V-shaped mesenteric defect), type IIIb ("apple peel" Atresia), type IV (multiple atresias) [7-11]. The type IV is seen in 1/5,000 to 1/4,000 live births. There is no gender predominance. Over a third of affected children are born prematurely. Less than 10% of jejunoileal atresia (JIA) are associated with extra-abdominal anomalies [12,13] in the postnatal period the clinical symptoms of this condition are that of neonatal bowel obstruction and the diagnosis is made with the

help of abdominal x-ray. However, abdominal ultrasound can help for antenatal diagnosis (30-50% of cases can be diagnosed before birth) [14-16].

Depending on the time to diagnosis and treatment, the type of atresia and associated conditions, the prognosis of JIA can be very good with over 90% of survival rate. Mortality varies from less than 10% in developed countries to more than 50% in low income countries [5,14]. The high survival rate in developed countries is due to advances in pediatric anesthesia, neonatal ICU, nutrition support including parenteral feeding. Survival has gradually improved to over 90% in developed countries [7]. These advances in management of JIA are lacking in low-income settings making the management of this condition challenging with high mortality rate [17].

# **Case Report**

We admitted in emergency a female newborn who was referred to us 7 days after birth from a primary health facility located on the outskirts of the city. Mother's history of pregnancy, labor and vaginal delivery was uneventful. The child was born mature weighing 3000g, Apgar score of 8/8/10. We didn't receive clear report from the primary health facility regarding patient progress and management. On admission, day 7 after birth, the child discharged meconium and the weight was 2300g, her general condition was worsening without any fever, the child was dehydrated and asthenic; she had abdominal meteorism and bilious vomiting. SPO2 95 %, HR 145bpm, RR 40 cpm, Temp 36°C; urine output was preserve but not quantified.

We noted upper abdominal bloating predominant in the epigastric and right hypochondrium region. The abdomen was supple with normal bowel peristalsis. No other anomaly was found. The anus was not obstructed. Plain abdominal x-ray showed distended loops of bowel greater than 3cm, collapsed colon, differential air-fluid levels, and thickened bowel wall (Figure 1). Prior to surgical exploration clinical diagnosis of small intestine bowel obstruction was made with presumption of pyloric stenosis and duodenal atresia. Blood exams showed Hb 13g%, Hematocrit 40%, Blood group O Rh +, Bleeding time 1 minute, Clotting time 3 minutes 30 seconds. We didn't do CBC, electrolytes, Urea, Creatinine.



Figure 1: Plain abdominal X-ray.

We secured an intravenous line for fluid resuscitation and protected the child against coldness. A number 6 nasogastric tube was placed preoperatively and maintained intraoperatively and postoperatively. The same applies to the n°5 pediatric Foley catheter. We administered 80 ml of secured blood, 100 ml of Normal Saline, 100 ml of 5% glucose serum and 50 ml of Ringer's lactate, IV Claforan 230mg BD, IM Gentamycin 3mg/kg OD, IV Paracetamol 20mg/kg.

After pre-anesthetic assessment we performed open laparotomy under general anesthesia. We found intraoperatively an important distension of the stomach and proximal jejunum; fibrous segment between narrowed segments approximately 60 to 70 cm from the ileocolic junction, extending to the proximal ileum ending in culde-sac. The distal segment of the ileum drained normally into the caecum. We confirmed the diagnosis of type IV jejunal atresia (Figure 2).



Figure 2: Distended stomach and proximal jejunum.

We performed resection of the atretic segment including part of jejunum and ileum; lateral anastomosis of the proximal jejunum to the ileum after checking the permeability of the distal segment of the digestive tract using saline through a catheter (Figure 3). Approximately 50g of meconium was discharged through the anus during this checking. We cleaned the peritoneal cavity, leave abdominal drain and close the abdominal wall. The child recovered fully from anesthesia within 2 hours. However, he vomited brown liquid, develop anemia (Hb: 7gr%), SPO2 85% and moderate dehydration. Lacking pediatric ICU, we managed the child in the recovering room. The child received 20ml/kg of blood in two portions depending on the state of its anemia, oxygen 1L/h, Ringer Lactate 108ml/kg and 10% glucose serum 108ml/kg IV. From day 1 post-operative follow up was uneventful. On day 5 the patient passed stool spontaneously, NGT was removed and the child returned to breastfeeding. On day 12 post surgery the sutures were completely removed (Figure 4) and the child was admitted in pediatric unit for nutrition and weight monitoring.



Lateral anastomosis jejunoileale

Figure 3: Lateral anastomosis of the proximal jejunum to the ileum.



Figure 4: Wound healing on day 12 post-operative.

# Discussion

Jejunoileal atresia (JIA) is a rare malformation in pediatric surgery unit, but it is the most common cause of neonatal bowel obstruction without gender predominance [12]. Boume et al. [18] reported an annual incidence of 2.1 cases over 10 years in Togo; a mean age of 11.2 days on admission with extremes of age between 2 and 28 days.

In our case, we have managed JIA in a 7-day female neonate. In sub-Saharan Africa, there is a frequent delay in receiving appropriate care. This is due to home births, religious and ancestral beliefs, and poor health system [14,19-21]. In India, on the other hand, early admission is the rule, and sometimes the diagnosis has already been made by antenatal ultrasound [22].

Our patient was born in a health center on the outskirts of the city and remained undiagnosed until the seventh day of life. As observed by Hounnou et al. [19], the delay of diagnosis in our case can be explained by the poverty of the population and the absence of health insurance companies, the lack of training of medical staff responsible for prenatal consultations, the under-equipment of hospital facilities and an inefficient referral system.

Boume et al. [18] reported a predominance of JIA cases in fullterm newborns (76%) compared with 23% of cases in premature newborns. We have managed a full-term newborn in our case. CHIKHI et al. [8] and Touloukian et al. [23] reported more than one third of JIA in premature children. Low birth weight is a poor prognostic factor [24]. HOUNNOU et al. [19] reported a mean birth weight of 2,648 + 152 g with extremes of 1,430 g and 3,530 g in newborn with JIA. We report a case of JIA in a newborn with 3000g birth weight. This may have contributed to our success. It appears from the literature that antenatal diagnosis is possible by means of an ultrasound scan carried out in the third trimester of pregnancy [15]. The advantage of antenatal diagnosis is the speed of surgical management in the immediate postnatal period, as this is an extreme emergency [13]. In addition, it can facilitate the choice of delivery route and rapid transfer to pediatric surgeon. In our case the patient's mother did not undergo any ultrasound scan during pregnancy. We believe that the socio-economic level of the mother, the under-equipment of the health center where she gave birth, poor referral system and the lack of training of the nursing and medical staff are all factors that could explain this.

In the postnatal period, jejunoileal atresia is clinically manifested by a neonatal occlusive syndrome. This clinical picture combined with plain abdominal x-ray make the diagnosis easy [19]. Guided by these tools, we were able to make the presumptive diagnosis and perform emergency surgery. JIA was confirmed intraoperatively. Maoneo A et al. [14] support that errors in clinical diagnosis of JIA are common and this is related to the lack of antenatal ultrasound, MRI and qualified personnel, making the diagnosis possible most of the time intraoperatively.

The gold standard of surgical treatment is resection and anastomosis [5,6]. We found the blind ends of intestine were separated by a defect in the intestinal blood supply; Distended stomach and proximal jejunum, presence of fibrous segments between narrowed segments extending to the proximal ileum which terminates in a cul-de-sac. About 20 cm of the small intestine was atretic. We classified it as type IV. We performed resection and lateral anastomosis. The immediate postoperative period was characterized by vomiting, anemia and low saturation.

Our patient needed pediatric ICU before and after surgery. We don't have one in our department; however, we have been able to conduct preoperative and postoperative resuscitation in the neonate room. Post-operative follow up is an important time during management of JIA especially those diagnosed late. MONEO et al reported a case of JIA who died from dehydration on day 2 post-operative [14]. On day 12, the sutures were completely removed and the patient continued management of the nutrional status with the pediatrician. According to the literature, the prognosis depends on the type of atresia, the location and especially the length of the affected small bowel [15].

# Conclusions

Jejunoileal atresia is a medical and surgical emergency requiring prolonged intraoperative resuscitation. Management is multidisciplinary; good pregnancy monitoring and careful clinical examination can promote early detection and diagnosis with a view to appropriate and rapid treatment, even in low-income settings.

# **Conflicts of Interest**

The authors declare no conflicts of interest with regard to this article.

# **Authors' Contributions**

- Kanyinda F MD: substantial contribution to design and assembly, data collection, presentation and interpretation.

- Talona R MD: revised and participated in the final approval of the version to be published.
- Kasereka F MD: revision and participation in the final approval of the version to be published.
- M'toro J MD: revised and participated in the final approval of the version to be published.
- Lokangu P MD: revised and participated in the final approval of the version to be published.
- Wami T MD: reviewed and participated in the final approval of the version to be published.
- Lukwamirwe A MD: revision and participation in the final approval of the version to be published.
- Bwini A MD: review and participation in the final approval of the version to be published.
- Tshimbila K MD. PhD: reviewed, wrote the final version and participated in the final approval of the version to be published.

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