A 5½ month old girl with ascites

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Presentation of Case

Dr. Zannatul Ferdous Sonia: A 5½ month old girl, 2nd issue of non-consanguineous parents from Jamalpur immunized as per EPI schedule, presented at the outpatient department with the history of abdominal distension since 2 months of her age which gradually increased day by day (Figure 1). She also had a history of fever for 1 month which was low-grade, intermittent in nature but not documented and subsided after taking antipyretic. She had no history of cough, contact with tuberculosis patient, jaundice, pale stool, diarrhea, scanty micturition, abdominal trauma or any history of surgery. Her elder brother was healthy and there was no family history of such type illness.

She lived in a well-ventilated tin-shade pucca house. On examination, the patient was afebrile, anicteric and mildly pale. Other vital signs were within normal limit. The edema was absent. She was anthropometrically well and thriving. Bedside urinary albumin was nil. BCG mark was present. Per abdominal examination revealed ascites. There was no organomegaly. Other systemic examinations revealed normal findings.

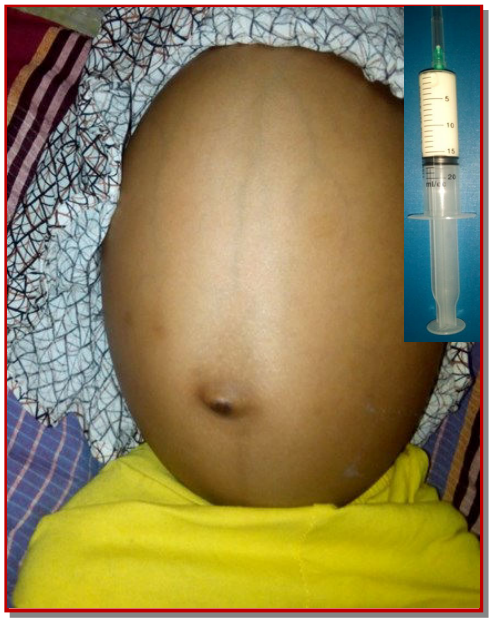

Figure 1: Abdominal distension with visible engorged vein, everted umbilicus and fullness of flank. Milky white ascitic fluid (inset)

For her complaints, she was admitted into the Mymensingh Medical College Hospital and underwent some investigations like complete blood count, serum T_3, T_4 and TSH, α-fetoprotein, alkaline phosphatase, Mantoux test, ultrasonography of the whole abdomen, etc (Table I). Her report showed mild anemia (hemoglobin 8.8 g/dL). The ultrasonography report showed ascites without organomegaly. She was then treated with vitamins and minerals and was referred to Dhaka for better management.

After admission in this hospital, blood was sent for serum alanine aminotransferase, serum triglyceride and chest X-ray was done (Table I).

Provisional Diagnosis

Congenital chylous ascites

Differential Diagnosis

Dr. Afsana Yasmin: As Bangladesh is a highly endemic zone for tuberculosis, so after evaluating the other features, it could be a case of abdominal tuberculosis (peritoneal type).

Abdominal tuberculosis

The prevalence of tuberculosis among the children in Bangladesh is 3%.[1] Childhood tuberculosis case detection is difficult due to the multifactorial problems such as low-quality of sampling, paucibacillary and lack of *Mycobacterium* culture facilities. In children, the number of extra-pulmonary tuberculosis cases are higher compared to the pulmonary tuberculosis.[1] The most common forms of abdominal tuberculosis in children are serous peritonitis and intestinal tuberculosis.[2] Tuberculous bacilli can reach the gastrointestinal tract by the following route: a) hematogenous spread from lung focus, b) ingestion of bacilli from infected sputum, c) unpasturized cow's milk ingestion, d) direct

Table I

Laboratory investigations of the patient

Parameter	Findings	References
Hemoglobin (g/dL)	8.8	13.5 ± 1.3
WBC count (×10⁹/L)	10	7.0 ± 3
Differentials		
Neutrophil (%)	52	40-80
Lymphocyte (%)	40	20-40
Eosinophil (%)	4	1-6
Monocyte (%)	2	2-10
Peripheral blood film	Microcytic hypo-chromic anemia	
Platelet count (10⁹/L)	365	150-400
Erythrocyte sedimentation rate (mm in 1st hour)	30	0-10
Serum T_3 (nmol/L)	2.6	1.2–3.6
Serum T_4 (nmol/L)	130.2	54–173
Serum TSH (μIU/mL)	2.0	0.3–5
α-Fetoprotein (ng/mL)	37.4	Up to 15.0
Alkaline phosphatase (U/L)	113	100–600
Mantoux test	4	>10
Ultrasonography of whole abdomen	Marked ascites	
Serum alanine transaminase (U/L)	20	0-55
Serum triglyceride (mg/dL)	222	<150
Chest X-ray	Normal	

spread from adjacent organ, e) lymphatic spread from adjacent lymph nodes.

Abdominal tuberculosis may be intestinal, mesenteric and peritoneal. The most common site of intestinal tuberculosis is the involvement of ileocecal region and peritoneal tuberculosis mainly presents with ascites.[3]

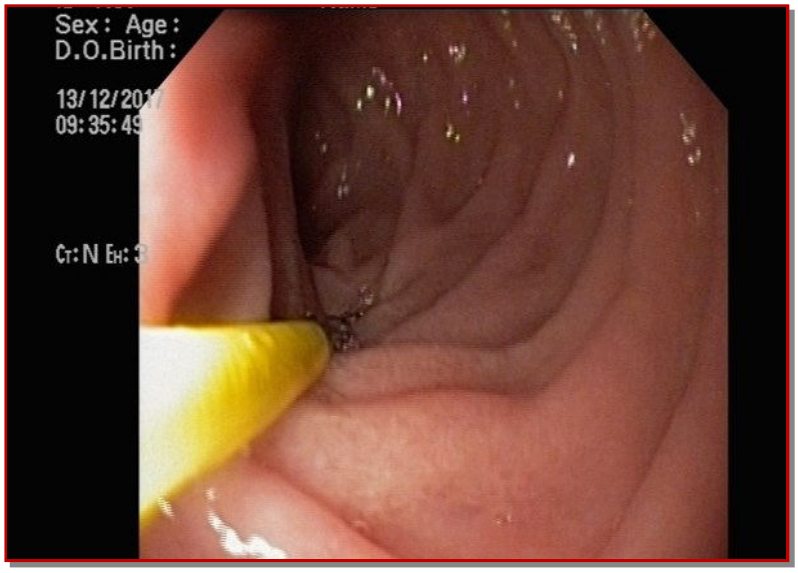

Figure 2: Endoscopy shows snowflaky spot on duodenum

Table II

Biochemical findings of ascitic fluid

Parameter	Findings
Appearance	Milky white
Cytology	Lymphocyte predominant
Protein (g/dL)	6.1
Triglyceride (mg/dL)	4329
Glucose (mg/dL)	95

The symptoms of abdominal tuberculosis are non-specific such as anorexia, malaise, fever, weight loss and abdominal distension which is due to ascites.

Physical examinations may reveal cachexia, fever, lymphadenopathy, absent BCG mark, hepatomegaly, splenomegaly, ascites, and features of intestinal obstruction.

Our patient presented with fever and ascites which are suggestive of peritoneal tuberculosis, but younger age, no history of cow's milk ingestion, no contact history with tuberculosis patient, absence of growth failure and no lymphadenopathy go against the peritoneal tuberculosis.

Omental cyst

Omental cyst is a rare condition which occurs usually during the early childhood and young adulthood. It is closely related to the mesenteric and retroperitoneal cysts and may be congenital or a benign neoplastic condition of lymphatic origin. Omental cyst usually presents with the abdominal distention of long duration, frequently with a palpable abdominal mass.[4] It may be unilocular or multilocular and contains fluid, which may be clear or straw-colored. There may be thick cheesy-white material which is due to the inspissated lymph fluid in the cyst.[5] In this patient, gradual abdominal distension since the early history goes in favor of congenital omental cyst but the presence of tense ascites and absence of palpable abdominal mass go against the diagnosis of omental cyst.

Dr. Luthfun Nahar: After evaluating the patient's presenting features, physical findings, initial investigation results, bedside paracentesis was done and ascitic fluid was found milky white in color (Figure 1). Ascitic fluid showed high concentration of triglyceride (Table II).

Dr. Wahiduzzzaman Mazumdar: I performed the endoscopy to identify the cause of chylous ascites. It showed snowflaky spots on second part of the duodenum (Figure 2). A biopsy was taken from the second part of duodenum and biopsy tissue was sent in formalin for histopathology. The histopatho-

logy report showed distorted villi, lamina propria contained a moderate number of lymphocytes and dilated lymphatics were present in the villous core with no granuloma or malignant cells; which were suggestive findings of intestinal lymphangiectasia.

According to history, physical examinations, investigations, endoscopy findings and histopathology report, it was a case of chylous ascites due to intestinal lymphangiectasia. There are many etiologies[6] which are responsible for chylous ascites.

Congenital lymphatic abnormality

The congenital lymphatic abnormality is the predominant cause of chylous ascites in the present case. It accounts for 84% of all causes in pediatric population.[6]

Intestinal lymphangiectasia

Intestinal lymphangiectasia is the pathological dilatation of the lymphatic vessels in mucosa and submucosa of the small intestine. It accounts for about 26% causes of chylous ascites in children and adolescent. Intestinal lymphangiectasia may be primary or secondary.

Neoplasia

It is another important acquired cause of chylous ascites. Among the malignancies, lymphoma accounts for at least one-third of the cases. Obstruction and invasion into the lymphatic channels due to malignancy leads to the disruption of normal lymphatic flow and causes chylous ascites.[6] There were no suggestive features of malignancy in this patient.

Tuberculosis

Tuberculosis of the gastrointestinal tract is the 6th most frequent site of extra-pulmonary tuberculosis. One manifestation of the abdominal tuberculosis is chylous ascites which is due to interruption and blockage of lymphatic drainage by enlarged lymph node.[10]

Filariasis

Filariasis, caused by the parasite *Wuchereria bancrofti*, may cause chylous ascites. This is due to the severe inflammatory reaction in the lymphatic vessels by parasites, leading to lymphedema and chylous ascites.[6]

Pancreatitis

Both acute and chronic pancreatitis can cause chylous ascites. Two mechanisms have been proposed for the development of chylous ascites in pancreatitis, which are compression of lymphatic channels or direct damage of lymphatic channel by the pancreatic enzymes in the peritoneal cavity.[6]

Trauma

Trauma or surgery cause direct leakage of the lymph through a lymphoperitoneal fistula into the peritoneal cavity due to acquired lymphatic disruption which cause chylous ascites.[6, 7] There is no history of trauma and surgery exclude the diagnosis.

Hemodynamic abnormalities

Constrictive pericarditis, right heart failure, dilated cardiomyopathy and rheumatic valvular disease can cause chylous ascites. The proposed mechanism is by increasing pressure within the lymphatic vessels.[8]

Cirrhosis

The underlying pathophysiology of chylous ascites is due to the rupture of serosal lymphatic channels, which are dilated due to excessive lymph flow. Other causes of portal hypertension, such as portal vein thrombosis, have also been implicated as a cause of chylous ascites.[9] As there was no history of jaundice and liver function tests were normal so these excluded the diagnosis of cirrhosis.

Though patient's age is now 5½ month and the abdominal distension started since 2 month of her age, she had no history of underlying liver, kidney or heart diseases and history was prolong with no significant weight loss, so this chylous ascites also was not due to malignancy. So, it is a case of congenital chylous ascites. Endoscopy and biopsy finding reports are suggestive for intestinal lymphangiectasia.

Dr. Nahar's Diagnosis

Chylous ascites due to congenital intestinal lymphangiectasia

Discussion

Dr. A. S. M. Bazlul Karim: Chylous ascites is defined as the extravasation of milky or creamy appearing peritoneal fluid rich in triglyceride, caused by the presence of thoracic or intestinal lymph in the abdominal cavity.[6]

It develops when there is a disruption of the lymphatic system, occurs due to traumatic injury or obstruction (from benign or malignant causes). Chylous ascites may be congenital or acquired.[6] In develop countries malignancy and abdominal surgery are common whereas in the developing countries tuberculosis is common acquired cause of chylous ascites.[10, 11] Abnormalities of the development and maturation of lymphatic vessels are the cause of primary or congenital process.[12, 13]

Intestinal lymphangiectasia may be primary or secondary. Primary intestinal lymphangiectasia occurs due to developmental abnormalities of the

lymphatic channel and secondary intestinal lymphangiectasia due to acquired disease such as malignancy, tuberculosis, filariasis, inflammatory bowel diseases, etc.

Primary intestinal lymphangiectasia is a rare disorder characterized by dilated intestinal lymphatic system resulting in lymph leakage into the small bowel lumen and responsible for chronic diarrhea, protein-losing enteropathy leading to lymphopenia, hypoalbuminemia and hypogammaglobulinemia.[14]

Primary intestinal lymphangiectasia is a common cause of congenital chylous ascites in children. The proposed mechanism is the exudation of lymph through the walls of retroperitoneal mega lymphatics into the peritoneal cavity which occurs with or without a visible fistula.[6] Patient of intestinal lymphangiectasia may also have low cholesterol and low calcium level. Ultrasonography, CT scan and barium X-ray can detect bowel thickening and edema.

Dr. Mazumder: Diagnosis of chylous ascites should be done by history taking, physical examination and relevant investigations. Abdominal paracentesis is the most important diagnostic tool for ascites. Depending on the clinical suspicion, ascitic fluid should be sent for cell count, culture, Gram stain, total protein, albumin, triglyceride level, glucose, ADA, lactate dehydrogenase, amylase, and cytology.[6] The presence of a milky, creamy appearing ascitic fluid with triglyceride content above 200 mg/dL is diagnostic for chylous ascites.[11]

Abnormal lymphatic drainage can be demonstrated by lymphoscintigraphy and it has no adverse effect.[6] However, lymphangiography is the gold standard to detect lymphatic obstruction. It has also a therapeutic role. But it has some adverse effects like contrast hypersensitivity, tissue necrosis, fat embolism, and even transient lymphedema and chylous ascites.[6]

Intestinal lymphangiectasia is confirmed by endoscopic observation of white viili/white spot/snow flake on duodenum and histologicaly presence of lacteal juice, dilated mucosal and submucosal lymphatic vessels with polyclonal normal plasma cells. Endoscopy may be negative when intestinal lesions are segmental or localized. In such cases, video capsule endoscopy is a useful tool to detect the presence of intestinal lymphangiectasia and to specify its localization.[14, 15] Laparoscopy and laparotomy can also be done to identify the traumatic or post-operative causes.

Primary chylous disorders are usually caused by congenital lymphangiectasia, with or without associated thoracic duct obstruction.[16]

In the present case, endoscopy showed snowflake spot on duodenum and histologically distorted villi with dilated lymphatics in villous core.

Dr. Sayma Rahman Munmun: What is the management plan of this patient?

Dr. Md. Rukunuzzaman: Nutritional, pharmacological and surgical therapies are available treatment options for the chylous ascites. Bowel rest followed by total parenteral nutrition (TPN) may be helpful. Dietary therapy with high-protein and low-fat diet with medium-chain triglyceride will reduce the chylous ascites.[17-20]

Pharmacotherapy as somatostatin and octreotide either alone or both in combination with TPN are effective in the management of chylous ascites due to different causes.[21] Orlistat, an inhibitor of gastric and pancreatic lipases, can minimize the ascites and triglyceride levels in the chylous ascites due to cirrhosis.[21] Paracentesis should be done to relieve respiratory distress and the replacement of albumin is not recommended unless the patient has cirrhosis.[6]

Surgical treatment as transjugular intrahepatic portosystemic shunt, laparotomy and fistula correction, peritoneovenous shunt, fibrin glue therapy are other treatment options for chylous ascites.

In this patient, dietary therapy and octreotide were given as treatment option for chylous ascites.

Dr. Parisa Marjan: What is the dose of octreotide to treat chylous ascites?

Dr. Sonia: Octreotide can be given by subcutaneous or continuous infusion. Subcutaneous infusion of 10 -40 µg/kg 12 hourly for a single day. Continuous infusion of 1-8 µg/kg/hours over 10 days. But many studies showed that continuous infusion with 0.5-2 µg/kg/hours has good efficacy, low cost and less adverse effects.[21-23]

In our patient, we started by intravenous infusion at a dose of 0.5 µg/kg/hours and gradually increased to 1 µg/kg/hours on day 3 and continued till day 7 then tapered and discontinued on day 10 (Figure 3).

Dr. Md. Benzamine: What is the mechanism of action of octreotide to reduce chylous ascites??

Dr. Nahar: Octreotide is an analog of somatostatin which has a longer duration of action. It has a wide spectrum of inhibitory effects on the anterior pituitary function, pancreas, endocrine secretions, and gastrointestinal tract. It inhibits the secretion of pancreatic hormones like insulin, glucagon, gastrin and other peptides, as a result, reduces pancreatic juice and also suppress gastric, pancreatic and small bowel secretion, and increases water and electrolyte re-absorption. It also reduces splanchnic blood flow, all these help to reduce ascites.[24, 25]

It is a pharmacologic agent that also augments the closure of any leakage in the lymphatic system.[26]

Dr. Mohua Mondol: What are the adverse effect of octreotide? How will you follow-up?

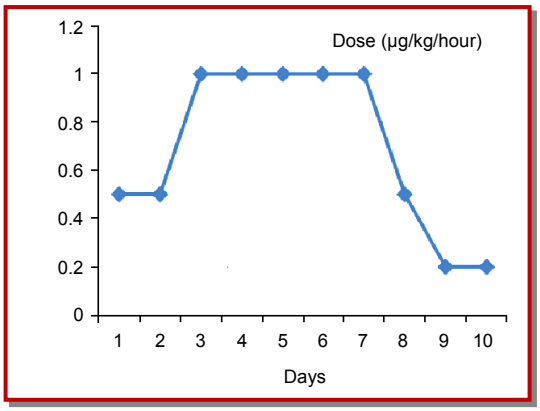

Figure 3: Dose and duration of treatment by octreotide

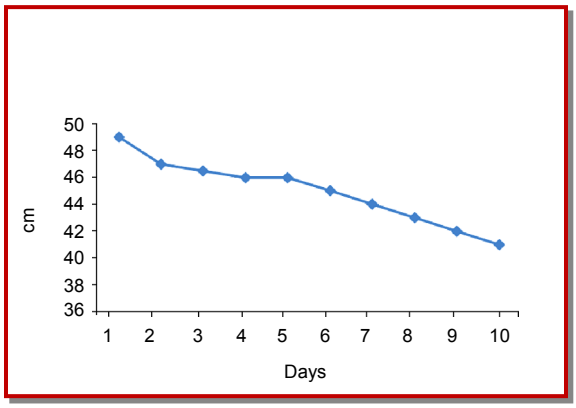

Figure 4: Abdominal girth (cm) after starting octreotide

Dr. Karim: Octreotide causes nausea, abdominal cramps, diarrhea, constipation, malabsorption of fat and flatulence. It can also cause hypertension, hypotension, hyperglycemia, hypoglycemia, cholestasis, hypothyroidism, etc.[24] Follow-up will be done based on clinical, laboratory and ultrasonography findings.[27]

Dr. Yasmin: How long the dietary management will be continued?

Dr. Sonia: The diagnosis of this patient is congenital intestinal lymphangiectasia, though it is not an acquired cause. So, the dietary management will be lifelong.

Dr. Kamrun Nahar: What is the prognosis of this patient?

Dr. Nahar: Abdominal girth was reduced after starting octreotide therapy (Figure 4).

Primary intestinal lymphangiectasia is a chronic disorder requiring long-term dietary control based on low fat with supplementation of medium chain triglyceride. It is rarely cured but can remain in remission for a long time. It is fatal when complications like pleural effusion and pericardical effusion occur and unresponsive to treatment.[14]

Final Diagnosis

Chylous ascites due to congenital intestinal lymphangiectasia

References

1. Islam Z, Sanin KI, Ahmed T. Improving case detection of tuberculosis among children in Bangladesh: Lessons learned through an implementation research. BMC Public Health, 2017; 17: 131.
2. Tinsa F, Essaddam L, Fitouri Z, Brini I, Douira W, Becher SB, Boussetta K, Bousnina S. Abdominal tuberculosis in children. J Pediatr Gastroenterol Nut. 2010; 50: 634-38.
3. Sharma MP, Bhatia V. Abdominal tuberculosis. Indian J Med Res. 2004; 120: 305-15.
4. Gary D. Shackelford GD, McAlister WH. Cysts of the omentum. Pediatr Radiol. 1975; 3: 152-55.
5. Ibrahim E. Bassiouny IE, Abbas TO. Omental cysts in children: Rare causes of abdominal masses (a report of two cases): Open J Pediatr. 2013; 3: 418-21.
6. Al-Busafi SA, Ghali P, Deschênes M, Wong P. Chylous ascites: Evaluation and management. ISRN Hepatol. 2014; 2014.
7. Gaag NA, Verhaar AC, Haverkort EB, Busch OR, Gulik TM, Gouma DJ. Chylous ascites after pancreaticoduodenoctomy: Induction of a grading system: J Am College Sur. 2008; 207: 751–57.
8. Lee YD, Lee YH, Choi HS. Chylous ascites in a patient with sepsis caused by bilateral pneumonia. Korean J Critical Care Med. 2014; 29: 217-21.
9. Tsauo J, Shin JH, Han K, Yoon HK, Ko GY, Ko HK, Gwon DI. Transjugular intrahepatic portosystemic shunt for the treatment of chylothorax and chylous ascites in cirrhosis: A case report and systematic review of the literature. J Vascular Int Radiol. 2016; 27: 112-16.
10. Barman B, Ete T, Warjri SB, Beyong T, Lynrah KG, Nobin H, Singh V. Chylous ascites and tuberculosis: A case report with literature review. Case Study Case Rep. 2015; 5: 74-80.
11. Shiba H, Wakiyama S, Gocho T, Ishida Y, Misawa T, Yanaga K. A case of successful conservative treatment for chylous ascites after living-donor liver transplantation. Int Surg. 2012; 97: 360-62.
12. Steinemann DC, Dindo D, Clavien PA, Nocito A. Atraumatic chylous ascites: Systematic review on symptoms and causes. J Am College Sur. 2011; 212: 899-905.
13. Kita RJ, Kornacka BMK, Dobrzañska A, Rudziñska I, Kowalska JC, Wawrzoniak T. Congenital chylous ascites. Polish J Radiol. 2011; 76: 58-61.
14. Vignes S, Bellanger J. Primary intestinal lymphangiectasia (Waldmann's disease). Orphanet J Rare Dis. 2008; 3: 5.

15. Mistilis SP, Skyring AP. Intestinal lymphangiectasia: The therapeutic effect of lymph venous anastomosis. Am J Med. 1966; 40: 634–41.

16. Kohnoe S, Takahashi I, Kawanaka H , Mori H, Okadome K, Sugimachi K. Combination of preoperative lymphangiography using lipiodol and intraoperative lymphangiography using Evans Blue facilitates the accurate identification of postoperative chylous fistulas. Surg Today. 1993; 23: 929-31.

17. Bhardwaj R, Vaziri H, Gautam A, Ballesteros E, Karimeddini D, Wu GY. Chylous Ascites: A review of pathogenesis, diagnosis and treatment. J Clin Transl Hepatol. 2017; 6: 1-9.

18. Smoke A, DeLegge MH. Chyle leaks: Consensus on management? Nut Clin Practice. 2008; 23: 529-32.

19. Martinez MS, Massie J. Chylothorax: Diagnosis and management in children. Pediatr Resp Riv. 2009, 10: 199-207.

20. Parrish CR, McCray S. When chyle leaks: Nutrition management options: Practical Gastroenterol. 2004; 28: 60-76.

21. Huang Y, Zhuang S, Li Y, Liu M, Chen H, Du M. Successful management of congenital chylous ascites in a premature infant using somatostatin analogue. Indian J Pediatr. 2011; 78: 345-47.

22. Pessotti CFX, Jatene IB, Buononato PEU, Elias PF, Pinto ACD, Kok MFJ. Use of octreotide in the treatment of chylothorax and chyloperitoneum. Arquivos Brasileiros de Cardiologia. 2011; 97: 33-36.

23. Talluri SK, Nuthakki H, Tadakamalla A, Talluri J, Besur S. Chylous ascites. North Am J Med Sci. 2011; 3: 438-40.

24. Philippe CP, Timsit J, Harris AJ. Clinical pharmaco -kinetics of octreotide. Clin Pharmacokinet. 1993; 25: 375-91.

25. Wilcock A, Twycross R. Therapeutic reviews. J Pain Symptom Manage. 2010; 40: 789-95.

26. Nurullah M, Karim ASB, Rukunuzzaman M, Mazumder MW, Begum F. Management of chylous ascites with octreotide in a Bangladeshi child: A case report. Bangladesh J Child Health. 2016; 40: 49 -51.

27. Campisi C, Bellini C, Eretta C, Zilli A, da Rin E, Davini D, Bonioli E, Boccardo F. Diagnosis and management of primary chylous ascites. J Vascular Surg. 2006; 43: 1244-48.