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Care of Child with Congential Microgastria

Esther Kanthi. K
(Lecturer, Paediatric Nursing, College of Nursing, Christian Medical College, Vellore, India)
Corresponding Author: Esther Kanthi. K

Abstract:
Congenital microgastria is an extremely rare anomaly of the stomach that occurs as a result of impairment of normal foregut development. It was first described in 1894, and to date, only 63 cases have been reported. It often occurs in a constellation of other anomalies of the gastrointestinal, cardiopulmonary, renal, central nervous systems and limb. Rarely does it occur in isolation. Congenital Microgastria presents with frequent vomiting, gastroesophageal reflux disease, recurrent aspiration pneumonia, and failure to grow. The diagnosis is made with a contrast study of the upper gastrointestinal tract which shows a small, tubular stomach in the midline, incompetence of the cardiac sphincter, and megaesophagus. Upper gastrointestinal scopy shows a dilated esophagus with a small, tubular or blind stomach. Treatment options include conservative management with frequent nasogastric tube feeding of small amounts of food and definitive surgical management by gastric augmentation with a jejunal loop known as Hunt–Lawrence procedure and total gastric dissociation with a Roux-en-Y esophagojejunostomy. Nursing care preoperatively is aimed at improving the nutritional status by administering tube feeding and positioning the child in fowlers to prevent aspiration. Postoperatively administer oxygen if required, intravenous fluid and antibiotic administration, pain management, initiation of oral feeds, prevention of complication and preparation for discharge by teaching about home care, continuation of medication and follow up.
I here present a child with congenital microgastria and have discussed the condition including the definition, associated anomalies, embryology, etiology, clinical manifestation, diagnostic evaluation, medical, surgical, and nursing management.

Keywords: *Congenital Microgastria, Gastroesophageal Reflux, Megaesophagus, Gastric Augmentation, Hunt-Lawrence Pouch*

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I. Introduction:

Congenital microgastria is an extremely rare anomaly of the stomach resulting from impairment of normal foregut development (1). It is characterized by a small, tubular stomach with a dilated esophagus and incompetence of the cardia (2). Congenital microgastria was first reported in 1894 by Dide in an adult and 1956 by Caffey in a child (3). It occurs either in isolation or is associated with a spectrum of anomalies of the gastrointestinal system such as asplenia, intestinal malrotation, tracheoesophageal fistula, esophageal atresia, pyloric atresia, duodenal atresia, Hirschsprung's disease, imperforate anus, transverse liver; anomalies of the cardiopulmonary system such as atrioventricular septal defects, laryngotracheobronchial clefts; renal anomalies such as renal dysplasia, renal aplasia; anomalies of the central nervous system such as hydrocephalus, agenesis of the corpus callosum; lumbosacral vertebral anomalies, micrognathia, anophthalmia and upper limb anomalies such as radial, ulnar, and thenar hypoplasia (1,4–8). Only 63 cases of microgastria have been reported in the literature until 2019 (9). It is seldom an isolated anomaly and just 5 such cases have been reported till 2011 (10).

Embryology:

The stomach originates as a dilatation of the foregut during the 4th week of embryological development. In the 5th week, the stomach achieves its normal contour through differentiated growth along its longitudinal and anteroposterior axis. The stomach starts as a straight tube but since there is differential growth of its dorsal and ventral wall, the greater curvature becomes more elongated than the lesser curvature. Continued differential expansion of the superior part of the greater curvature gives rise to the fundus and cardiac incisura by the end of the seventh week. During the seventh and eighth weeks, the stomach rotates 90° around a cranio-caudal axis. The former anterior portion will form the right side of the stomach and the former posterior portion becomes the left side of the stomach. Early arrest in gastric development results in microgastria with an impaired reservoir capacity (11–13).

Etiology:

Congenital microgastria is an idiopathic disorder, where the diminutive stomach lies in the midsagittal plane and does not develop a fundus, corpus, or antrum (1). It is postulated that congenital microgastria results from arrested gastric development during the first trimester of prenatal life (2). The stomach and the spleen are both derived from the embryonic dorsal mesogastrium; hence the association with asplenia is common. The association of limb, cardiac, tracheoesophageal, vertebral, and renal anomalies with congenital microgastria has been attributed to impairment of early mesodermal development, most probably due to gene dysfunction (11). It has been suggested that microgastria in association with limb reduction defects and central nervous system anomalies has an autosomal recessive pattern of inheritance (7).

Clinical manifestations:

The clinical manifestations vary significantly depending on the phase and degree to which stomach maturation is arrested. Children with congenital microgastria present with postprandial vomiting, recurrent aspiration pneumonia, feeding difficulty, failure to thrive, growth retardation, and malnutrition secondary to the small stomach and the associated gastroesophageal reflux (1,2,4).

Diagnosis:

- **Antenatal ultra-sonogram:** Fetal stomach is visualized after 9 weeks of gestational age in antenatal ultrasonography (14). Prenatally microgastria mimics esophageal atresia with non-visualization of stomach and polyhydramnios (15).
- **Upper gastrointestinal barium study:** Shows a narrow, fine tube in the midline, frequently associated with the incompetence of the lower esophageal sphincter and consequently megaesophagus. The esophagus dilates to compensate for the absence of the stomach (11).
- **Upper Gastrointestinal scopy:** Shows dilated esophagus with a small, tubular or blind stomach
- **Roentgenographic study:** Shows an esophagus dilated proximally and narrowed at the apparent cardioesophageal junction and a tubular stomach, without recognizable differentiation into fundus, body, antrum, and pylorus (4).
- **Stomach Biopsy:** Several studies have shown normal gastric mucosa; however the total cell mass of the stomach is severely reduced, leading to reduced production of acid and intrinsic factor (11).

II. Management:

In patients with lesser degrees of microgastria, conservative nonsurgical treatment with small frequent amounts of oral or nasogastric feedings, positioning precautions, and prokinetic medications are found to be effective. Small, frequent feeding has although had good results (2,16), frequent small feeds or a gastrojejunostomy to drain the stomach causes uniform poor somatic growth, sexual underdevelopment, delay in cognitive milestones, failure to thrive, and dumping syndrome (5,11). Other surgical interventions such as gastrostomy and gastroenterostomy, have also not been successful because of continued gastroesophageal reflux (16,17).

In patients with more severe dysgenesis, a conservative approach typically is unsuccessful. Bypass or feeding jejunostomy has been reported for cases with long-term survival (12). Gastric augmentation by creating a Hunt-Lawrence (HL) double-limb jejunal pouch is used to enlarge gastric reservoir capacity. It was first described by Neifeld et al in 1980 (2,17). The HL pouch is created by making a Roux-en-Y loop of the jejunum, joining the afferent and efferent loops together to prevent alkaline reflux, and a food pouch, made by anastomosing a folded loop of a distal jejunal segment to the stomach (5,18). The pouch increases the size of the gastric reservoir, thereby allowing the resolution of the secondary esophageal changes and permitting normal growth to proceed (16). The HL pouch has excellent results relieving symptoms of reflux and facilitates nutritional management (12,19). Prophylactic penicillin is essential for patients with asplenia (19). In the postoperative period, patients with microgastria require careful follow-up and chronic administration of vitamin B 12 (16,19).

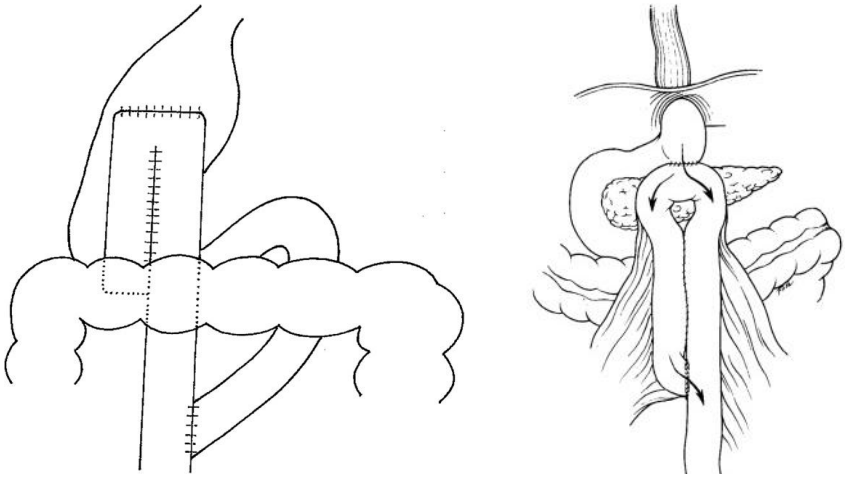

Fig 1: Line diagram showing the HL procedure for gastric augmentation. The afferent and efferent limbs of a Roux-en-Y loop of jejunum are anastomosed together and joined to the microstomach.

Complications:
Post HL pouch, postprandial symptoms after total gastrectomy, such as dumping syndrome, epigastric pain, steatorrhea, fat malabsorption, and bacterial overgrowth, have been reported (19,20).

Prognosis:
The long-term quality of life and somatic growth after surgery, however, are not well known (5).

Nursing management:
The nursing management of a child with congenital microgastria is presented using a case report

III. Case Report:

A 6 year old male child, Master A, presented with complaints of persistent non-bilious vomiting, poor feeding, failure to thrive and recurrent aspiration pneumonia since 1 year of life. He also complained of epigastric pain, heart burn, loss of appetite and loss of weight. He was born at term, to a primi gravida mother who had no prenatal complications. The child birth was by normal vaginal delivery, conducted by trained personnel in rural hospital, India, in 2010. The baby was breast fed soon after birth. At 1 year of age, he presented with complaints of vomiting and difficulty in swallowing to a hospital, where he was diagnosed to have hypertrophic pyloric stenosis and he underwent pyloromyotomy in December, 2011. He continued to have the symptoms post pyloromyotomy and was evaluated for the same. He was then diagnosed to have achalasia cardia and underwent Heller's cardiomyotomy in April, 2012.

Since he continued to have the symptoms such as progressive dysphagia for solids and liquids, epigastric pain, feeding and failure to thrive subsequent to the surgeries he was brought here. On evaluation, Barium swallow demonstrated dilated esophagus with mild narrowing at the gastroesophageal junction with reflux of contrast from the fundus of the stomach into the esophagus (Fig 2). The rest of the stomach distally and the duodenum was not opacified. Therefore he underwent feeding jejunostomy to improve his nutritional status in December, 2016.

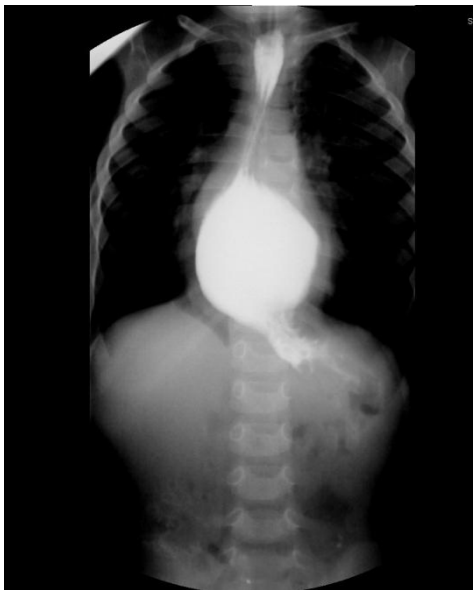

Fig 2: Barium swallow showing dilated esophagus with mild narrowing at the gastroesophageal junction with reflux of contrast from the fundus of the stomach into the esophagus

After 8 months, he came back with complaints of not taking anything orally and his nutritional status continued to be poor. He was therefore admitted to improve his nutritional status and for further evaluation. On assessment, child was pale and undernourished. Upper Gastrointestinal (GI) scopy showed dilated esophagus with nodularity, coated with whitish plaque suggestive of fungal esophagitis and small blind stomach. Gastrografin swallow study showed dilated esophagus with mild narrowing at the gastroesophageal junction with reflux of contrast from the fundus to the esophagus (Fig 3). The rest of the stomach distally and duodenum was not opacified. Therefore the child underwent Hunt Lawrence J pouch, where the feeding jejunostomy was taken down and J pouch was made from jejunum and Roux-en-Y anastomosis was done. Nasojejunal (NJ) tube was placed beyond the anastomosis. Small NJ feeds was initiated on the 3rd postoperative (POP) day, oral liquids on the 8th POP day and the child was taking normal diet by the 13th POP day. At discharge he was recommended Vitamin B12, Folic acid and multivitamin supplements.

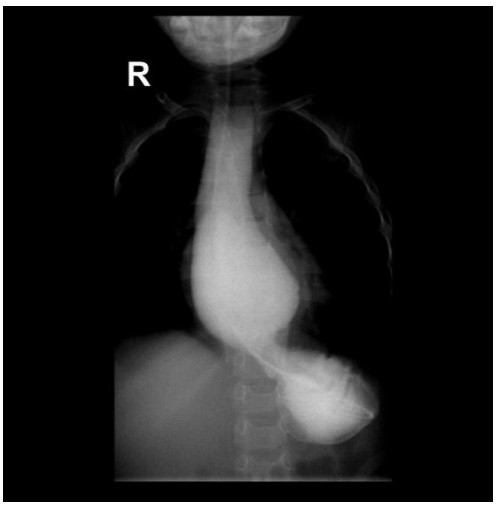

Fig 3: Gastrografin swallow showing dilated esophagus with mild narrowing at the gastroesophageal junction with reflux of contrast from the fundus to the esophagus

Nursing care (21):

Preoperatively, the child and the parents were counselled for fear, anxiety, knowledge deficit and nutrition. Measures were taken to prevent infection. The important preoperative and postoperative care is presented below based on the needs and problems of the child.

Preoperative Nursing diagnosis:
Imbalanced nutrition: less than body requirement related to inability to take enough food due to reflux, heart burn, vomiting as evidenced by inadequate food intake, weight loss, vomiting, regurgitation and failure to thrive

Interventions:
- Assessed the child's weight daily
- Obtained the nutritional history of the child
- Encouraged oral intake of small frequent meals that are easier to digest
- Administered high calorie and high protein foods through the feeding jejunostomy
- Instructed parents to nurse in upright position at least 2 hours after meals, avoid eating 3 hours before bedtime
- Instructed patient to eat slowly
- Encouraged to avoid food or drink 2 hours before bedtime or lying down after eating
- Elevated the head of the bed

Evaluation:
Optimal nutritional status was maintained as evidenced by absence of further loss of weight and normal laboratory parameters

Nursing diagnosis:
Risk for Aspiration related to esophageal compromise affecting the lower esophageal sphincter

Interventions:
- Assessed for pulmonary symptoms resulting from reflux of gastric content
- Assessed patient's ability to swallow and the presence of gag reflex
- Avoided placing patient in supine position and had the child sit upright after meals
- Instructed parents to avoid highly seasoned food, acidic juices, bedtime snacks, and foods high in fat
- Elevated head end of the bed
- Instruct the child to chew food thoroughly and eat slowly
- Administered high calorie and high protein foods through the feeding jejunostomy

Evaluation:
Aspiration was prevented as evidenced by no pulmonary signs of reflux of gastric content.

Postoperative Nursing diagnosis:
Risk for ineffective airway clearance related to pooling of secretions secondary to general anaesthesia

Interventions:
- Assessed airway patency by checking bilateral air entry and breath sounds and vital signs
- Suction apparatus was kept ready to be used if required
- Positioned the child in semi‑fowlers
- Assisted with ambulation from the 2nd post‑operative day
- Ensured adequate hydration to easily liquefy secretions
- Administered intravenous fluids as per order

Evaluation:
Patent airway was maintained as evidenced by normal breath sounds, normal rate and depth of respirations.

Nursing diagnosis:
Risk for ineffective breathing patterns related to the relaxation of smooth muscles secondary to general anaesthesia

Interventions:
- Assessed respiratory rate, rhythm, quality, bilateral chest movements and saturation
- Administered 4L/min oxygen per minute through face mask for first 6 hours postoperatively
- Positioned in semi‑fowler
- Encouraged deep breathing exercises
- Encouraged ambulation

Evaluation:
The child was able to maintain an effective breathing pattern, as evidenced by relaxed breathing at normal rate and depth and absence of dyspnoea.

Nursing diagnosis:
Risk for decreased cardiac output related to blood loss during surgery

Interventions:
- Monitored heart rate, blood pressure (BP), urine output, capillary refill and SpO2
- Observed for signs of bleeding or oozing
- Administered IV fluids
- Monitored haemoglobin, platelet count and clotting time
- Maintained intake‑output chart

Evaluation:
Adequate cardiac output was maintained as evidenced by BP and pulse rate and rhythm within normal parameters for patient, strong peripheral pulses and an ability to tolerate activity without symptoms of dyspnea.

Nursing diagnosis:
Acute pain related to surgical incision

Interventions:
- Assessed pain score, location and characteristics Q4H. Pain was initially 7 then later reduced to 3
- Monitored vital signs
- Child was on epidural analgesia till the 4th post‑operative day, along with this, SC injection Morphine was also administered
- Non‑pharmacological measures such as positioning and use of comfort devices helped in making the child pain free

Evaluation:
Pain was reduced as evidenced by decrease in pain score, normal vital signs

Nursing diagnosis:
Risk for imbalanced nutrition less than body requirement related to nausea, vomiting, Nil Per Oral (NPO) status and inadequate intake.

Interventions:
- Administered IV fluid according to the patient's needs
- Small NJ feeds was initiated on the 3rd postoperative (POP) day, oral liquids on the 8th POP day and the child was taking normal diet by the 13th POP day.
- Administered antiemetic (injection Ondansetron) Q8H
- Administered Vitamin B12, Folic acid and multivitamin supplements.
- Taught parents about giving small, frequent meals rather than large ones and to include proteins, Vitamin C, iron and fibre‑rich diet for better recovery.

Evaluation:
The child was able to maintain optimal nutritional status as evidenced by the absence of loss of weight and normal laboratory parameters

Nursing diagnosis:
Risk for delayed surgical recovery related to complications secondary to surgery:

Interventions:
Haemorrhage
- Monitored vital signs
- Assessed for any signs of abdominal pain or discomfort
- Assessed for bleeding/oozing, monitored haemoglobin levels
- Administered IV fluids or blood products as indicated.

Infection

- Monitored vital signs and for signs of infection
- Maintained aseptic techniques and hand hygiene strictly
- Changed dressings daily
- Ensured good personal hygiene and a clean environment Administer prophylactic antibiotics

Dumping syndrome

- Assessed for signs such as dizziness, diaphoresis and diarrhoea
- Advised small periodic meals and to lie in low fowlers for 20–30 min after diet intake
- Instructed to avoid taking fluids with diet and to take complex carbohydrates instead of simple ones

Evaluation:
The child did not develop any complications.

Nursing diagnosis:
Deficient knowledge regarding home care and follow‑up related to lack of information

Interventions:

- Assessed level of knowledge and understanding on the teachings previously given (post‑operative exercises, diet and pain management)
- Explained about regular follow‑up, medications, home care, prevention and detection of complications such as infection, incisional hernia and dumping syndrome
- Encouraged the parents to clarify doubts and reinforced the information.

Evaluation:
The parents verbalized understanding about all the instructions given to them.

IV. Conclusion:

Master A's postoperative was uneventful. He was started on NJ feeds on the 3rd POP day, oral liquids on the 8th POP day and normal diet on the 13th POP day. He was able to tolerate the feeds given to him. He was discharged on the 14th POP day with vitamin 12, folic acid and multivitamin supplements. At the time of discharge he was afebrile, tolerating oral feeds well and had a healthy surgical incision. Congenital microgastria is a rare anomaly and often is present with other life threatening anomalies. Timely identification and appropriate management will help patients to have an optimal nutritional status and prevent complications.

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