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RESEARCH ARTICLE

A RARE ASSOCIATION OF MORGAGNI HERNIA WITH DOWN SYNDROME.

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Abstract

Morgagni hernia is a rare diaphragmatic hernia accounting for only 2% of the congenital diaphragmatic defects. A case of Morgagni hernia was diagnosed radiologically in a 2 year old male with Down syndrome, with complaints of abdominal distension and not able to gain adequate weight ,with no respiratory distress. The 2-dimensional echocardiography was normal except patent foramen ovale right to left shunt. The diagnosis of Morgagni hernia was confirmed by Barium studies and Ultrasound Abdomen. The patient underwent a corrective surgery at 2 years of age. Literature review revealed only 18 cases of Morgagni hernia with Down syndrome reported till date, with age of presentation varying from neonatal age group to 12 years of age. The mode of presentations varied from asymptomatic detection to severe respiratory distress.

Introduction:-

Morgagni hernia is a rare diaphragmatic hernia accounting for only 2% of the congenital diaphragmatic defects. A case of Morgagni hernia was diagnosed radiologically in a 2 year old male with Down syndrome, with complaints of abdominal distension and not able to gain adequate weight ,with no respiratory distress. The 2-dimensional echocardiography was normal except patent foramen ovale right to left shunt. The diagnosis of Morgagni hernia was confirmed by barium studies and ultrasound abdomen. The patient underwent a corrective surgery at 2 years of age. Literature review revealed only 18 cases of Morgagni hernia with Down syndrome reported till date, with age of presentation varying from neonatal age group to 12 years of age. The mode of presentations varied from asymptomatic detection to severe respiratory distress.

A two-year-old male child, a product of a non-consanguineous marriage born of a singleton first pregnancy to a 23-year-old female with birth weight 3.1 kgs All his milestones were delayed. This child was born with clinical features of Down syndrome. This patient had required hospitalisation with complaints of abdominal distension and not able to gain adequate weight. Examination of the patient revealed features of Down syndrome (figure 1) -Mongoloid slant, flat facial features, low-set ears, focal alopecia in occiput, protruded tongue, bilateral clinodactyly, and generalised hypotonia ,muscle wasting and skin laxity. The electrocardiogram and 2-Dimensional echocardiography were normal except patent foramen ovale right to left shunt. Thyroid profile within normal limits. Analysis of the chest

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roentgenograph revealed a possibility of herniation of the bowel loops into the right hemithorax (figure-2) Ultrasonography of the chest showed presence of bowel loop in the chest. Barium enema revealed herniation of the bowel loop in the right hemithorax through a retrosternal- anterio medial defect confirming the presence of the morgagni hernia.

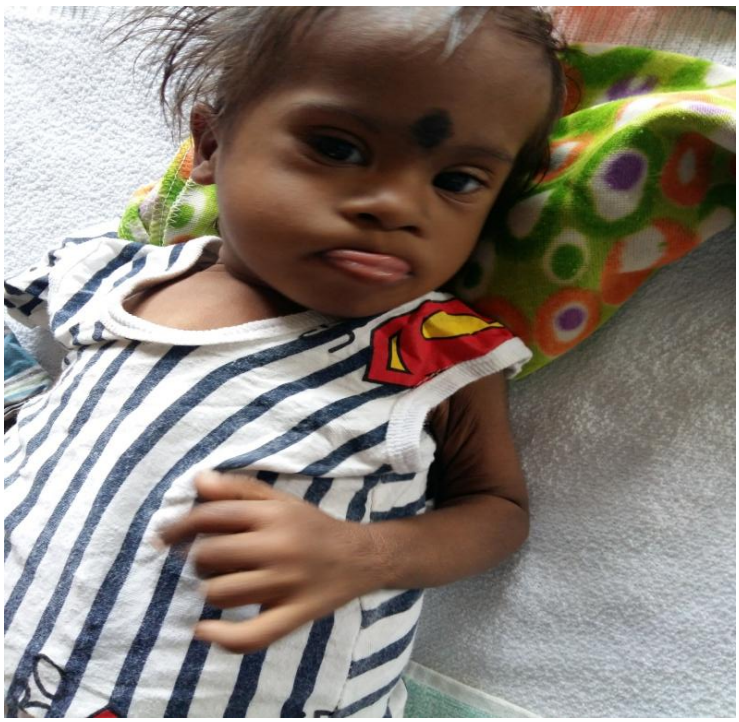

Figure 1:- Clinical Features of Down Syndrome.

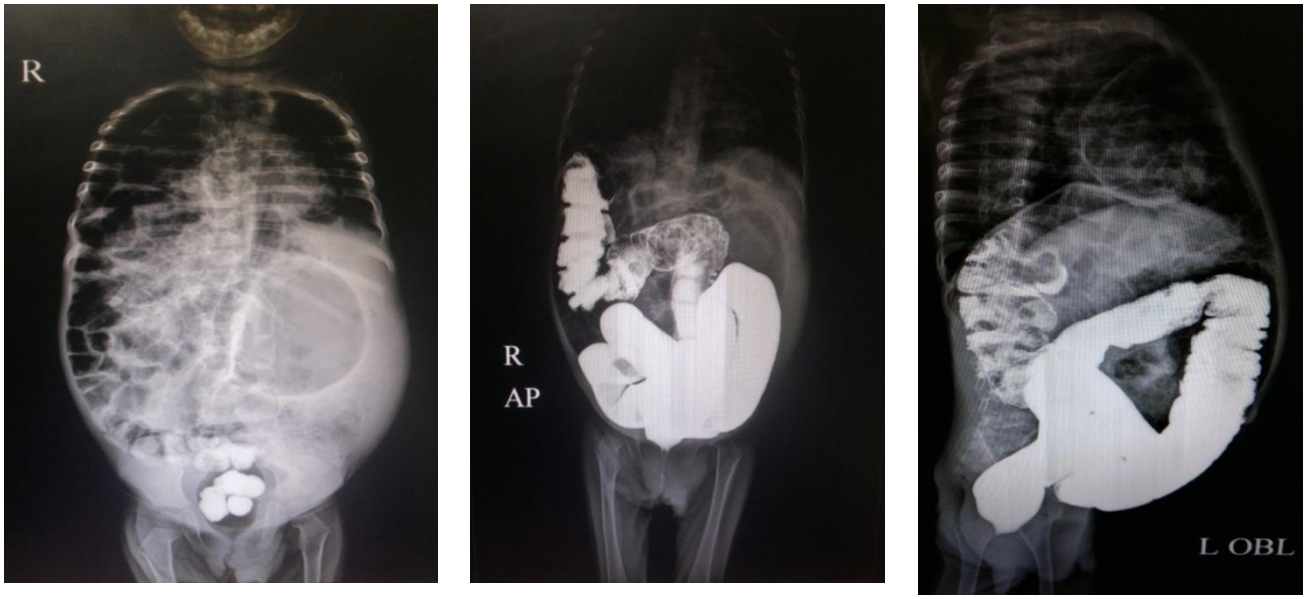

Figure 2:- X Ray Abdomen And Barium Enema

KARYOTYPING :47,XY+21 –DOWN SYNDROME (FULL FREE TRISOMY)

Pre operative weight 5.4 kg:-

The parents were counselled. The patient underwent a corrective surgery at the age of 2 years through transabdominal route-upper midline incision.Intra operatively,multiple lymphatic microcysts found in both in mesenteric and anti mesenteric border of small bowel and transverse colon(figure 3)microcyst biopsy taken sent for

HPE.Defect of size 5*4 cms noted in right antero medial region of diaphragm(figure 4).contents found to be small bowel was reduced following which the excess hernial sac was excised and the defect was closed with prolene in two layers from posterior lip of the defect to the periosteum of xiphisternum and ribs. There was no associated malrotation of the gut. Post Operative period was uneventful(figure 5).
POST OP HPE REVEALED LYMPH CYST.
Post operatively-2 weeks later ,significant weight gain-6.3kg

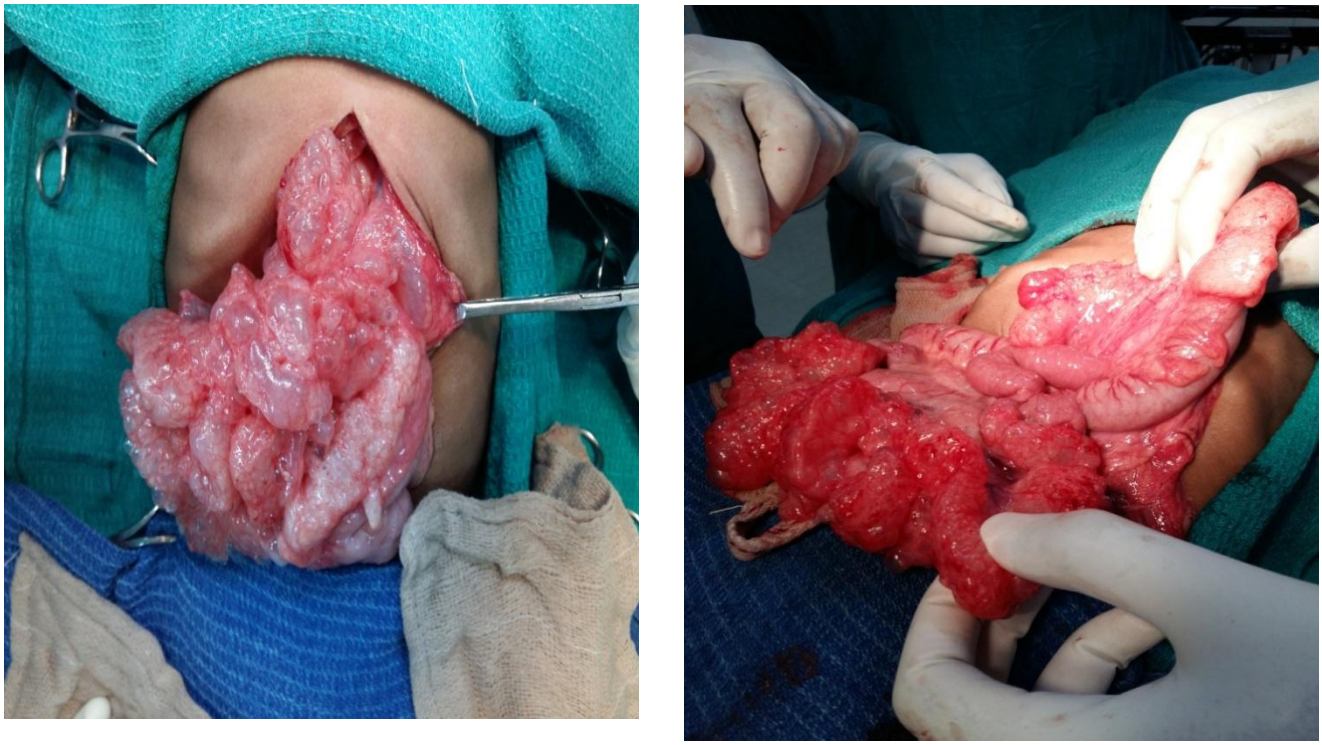

Figure 3:- Lymphatic Microcysts Seen In Small Bowel And Transverse Colon.

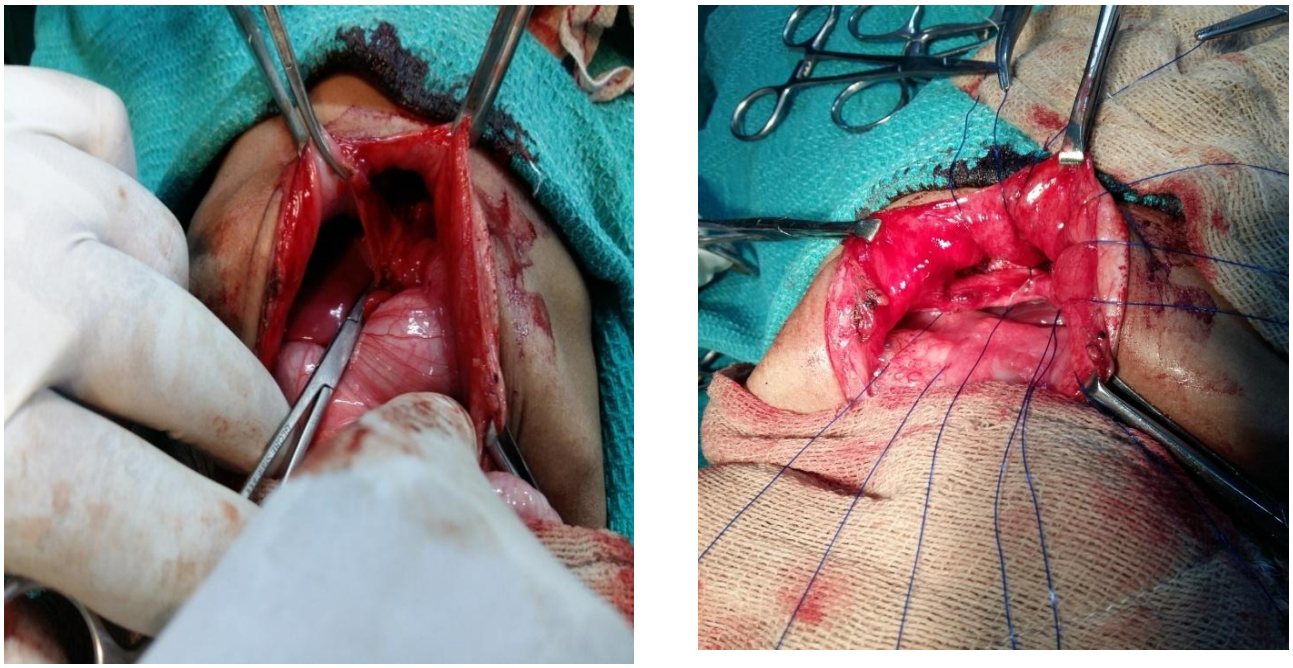

Figure 4:- Diaphragmatic Defect Of Size 5*4 Cms

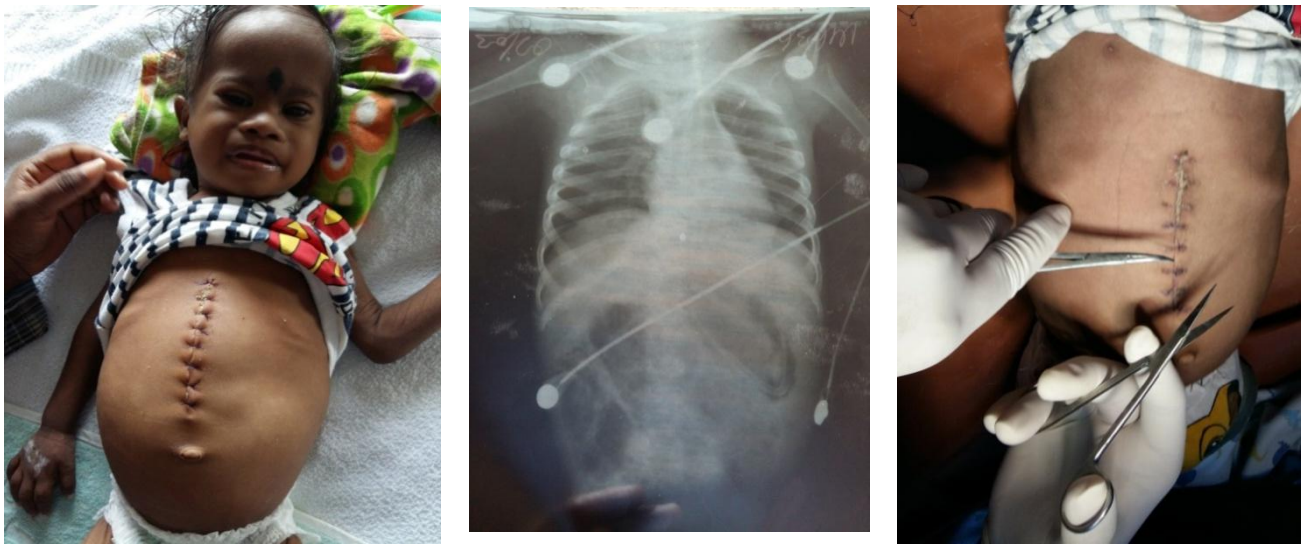

Figure 5:- Post Op Patient With Chest X Ray and suture removal after 2 weeks

Discussion:-

Morgagni hernia is the least common of the congenital diaphragmatic defects occurring with a varied frequency of 1 to 5.1% in a large series.[3],[12],[13] The foramen of Morgagni is a retrosternal space resulting from the failure of fusion of the fibrotendinous portion of the pars tendinalis arising from the costochondral arches with the fibrotendinous portion of the pars sternalis. This space is usually filled with fat and covered by pleura superiorly and peritoneum inferiorly. When present it offers a path through which abdominal viscera can herniate into the chest.

Foramen of Morgagni hernia occurs at the anteromedial portion of the junction of the thoracic wall and septum transversum. 90% of this occurs on the right side, 8% on the left and remaining are bilateral.[1]

The vast majority of the cases of Morgagni hernia are detected in asymptomatic children and adults.[3] Vague epigastric discomfort may be the only symptom in many cases. Rarely, the hernia presents in neonates & infants, in whom symptoms such as respiratory distress and cyanosis could be present. Like the Bochdalek hernias at a similar age, other manifestations include cough, choking episodes, vomiting following feeds, constipation, diarrhoea, failure to thrive, post-prandial fullness, and respiratory infections.[3],[5] Some cases may be discovered incidentally by visualizing the air-fluid levels or solid masses in the retrosternal region or on the right side of the chest radiograph.[5] Many cases may remain undetected and present only later in life. This may be due to the fact that though the defect is present from birth, presence of a sac resists herniation through the defect. Rupture of the sac by trauma or raised intra-abdominal pressure allows herniation and development of symptoms. Alternatively viscera may be herniated at an early stage, but patient remains asymptomatic till bowel complications occur.[5] The most common contents of the hernial sac are colon, small bowels, liver, omentum, and stomach.

So far literature search revealed only 18 cases of Down syndrome who had associated Morgagni hernia. The age of presentation varied from neonatal age group to 12 years of age.[7],[13] The mode of presentation varied from asymptomatic detection to the presentation with respiratory distress as in our case.[3],[14] Isolated case of retroperitoneal teratoma with Down syndrome and Morgagni hernia has also been described.[8] Interestingly, identical twins of Down syndrome with identical heart disease and Morgagni hernia have been reported.[7].

Although these cases, represent only a small percentage of patients with Morgagni hernia, this suggests that there may be a genetic component to Morgagni hernia too. This significant association between the hernia of Morgagni

and trisomy 21 may reflect defective dorsoventral migration of rhabdomyoblasts from the paraxial myotomes, caused by increased cellular adhesiveness in trisomy 21.[2].

This association has clinical implication too. In addition to screening for congenital heart disease, hypothyroidism, refractive errors; screening procedure for Morgagni hernia may also be undertaken especially in the cases of Down syndrome with respiratory manifestations. In cases of Morgagni hernia, one would have to be cautious to detect cases of mosaic Down that may be missed clinically.

Postnatally besides respiratory distress, clinical examination is usually unrewarding in Morgagni's hernia. Although chest radiograph may raise suspicions about the presence of Morgagni hernia, the diagnosis should be confirmed by barium studies, ultrasonography, computerised tomographic scan.[3],[5]Incarceration and strangulation are the complications of an unoperated hernia. Respiratory compromise due to compressed pulmonary parenchyma may necessitate an early surgery.[7] Surgical repair is usually undertaken through a transabdominal route but transthoracic approach is advocated, if the hernial sac has solid contents. Newer approaches like video-assisted thoracic surgery and laparoscopic repair hold promising future. The latter offers an additional diagnostic advantage as well as the potential for reducing morbidity when compared to laparotomy.[14]

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