# **Congenital Right Intra-Thoracic Hiatal Hernia**

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# **ABSTRACT:**

### **BACKGROUND:**

Congenital right sided intra-thoracic hiatal hernia is an uncommon clinical

entity consisting of herniation of a viscous through congenitally abnormal esophageal hiatus into an intra-thoracic location .

#### **AIM OF THE STUDY :**

Is to present the diagnostic challenge and the surgical management of eight cases of this uncommon congenital anomaly admitted during asix years period (2000 - 2005) to the medical city teaching complex in Baghdad. Iraq and to compare the study with other international studies. **METHODS**:

Eight patients were studied retrospectively in details as regard the age , sex , clinical manifestations , radiological findings . Details of the operative findings and the surgical outcome . **RESULTS :** 

All the patients had right sided intra-thoracic abdominal organ herniation mostly the stomach , all the patients ahad large hiatus , the patients were managed successfully through laparatomy with uneventful postoperative course .

#### **CONCLUSION:**

This uncommon variety of the congenital hiatal hernia presented a diagnostic challenge to the pediatrician , pediatric surgeon and the thoracic surgeon .Laparatomy was the procedure of choice and adding an anti –reflux procedure is highly indicated .

KEY WORDS: Hiatal Hernia, Gastro-esophageal reflux, Gastro-esophageal junction

#### **INTRODUCTION:**

Congenital hiatus hernia is the herniation of a viscous or part of it through the esophageal hiatus to an intra-thoracic location .

The ascent of the stomach into the chest through the esophageal hiatus is common and a poorly understood lesion .It is most oftenly situated in the midline or left hemithorax ; however a congenital right sided intra-thoracic hiatus hernia is uncommonly reported <sup>(1)</sup>.

Enlarged esophageal hiatus is common to sliding and Para esophageal hiatal hernia .The intrathoracic location of the esophago-gasric junction differentiates the more common sliding from the Para esophageal hiatal hernia .Symptomatic patient with a sliding hiatal hernia complains of persistent gasrtoesophageal reflux ;while symptoms in the paraesophageal hiatal hernia are mainly due to mechanical obstruction secondary to incarceration or volvulus <sup>(2)</sup>.

Congenital hiatal hernia constitutes (5-10 %) of all diaphragmatic defects . It is the abnormal hiatus rather than the congenital short esophagus which leads to the herniation of the stomach into the chest <sup>(3)</sup>.

\*College of Medicine , University of Baghdad \*Medical City Teaching Complex Al-Mansoor Pediatric Hospital Surgery is the only method of treatment in congenital hiatal hernia which involves reduction of the hernia , resection of the sac , repairing the hiatus and adding an anti-reflux procedure .

#### **MATERIALS AND METHODS:**

This is a retrospective and comparative study of eight children with congenital right sided intrathoracic HH which were admitted to the Medical City Teaching complex in (Baghdad –Iraq ) during a six years period between 1st. of January 2000 and the thirty  $1^{st}$ . of December 2005.

These cases were retrospectively studied as regard the age , gender , presenting symptoms ,radiological findings , surgical approach and the fruitfull outcome .

These children were carefully evaluated as regard history taking from their mothers , physical examination and for all of them plain chest X-Ray and contrast study were performed .The management was planned carefully in a team work between the thoracic and the pediatric surgeon.

Pre-operative bronchoscopy was done in two cases to clear the accumulated endo-bronchial secretion in order to reduce the possibility of post-operative chest infection and or atelectasis. All the children received pre-operative antibiotics which were mainly used to control chest infection and their mothers were instructed to give frequent small feeding and the maintenance of nearly upright posture during feeding in order to control the symptoms of reflux .

Surgical treatment was indicated in all the cases as all exhibited a major translocation of the stomach into the chest and the aim of surgery was to reduce the hernia , repair the hiatus , creating an intra-abdominal esophagus and the fixation of the stomach it its normal position below the diaphragm to prevent recurrence and to avoid future reflux with Nissen ' fundoplication ,where the gastric fundus was wrapped around the oesophagus.

In only one patient the pre-operative finding of delayed gastric emptying on barium meal necessitated preoperative pyloric myotomy.

The hiatus in all the cases was repaired using interrupted non absorbable suture material as 2/0 silk or 2/0 polyester and finally adding antireflux procedurePost-operative naso-gastric tube was maintained for forty –eight hours to avoid any gastric distension and nothing by mouth for the same period during which the patient is maintained on intra-venous fluid calculated according to the child weight and daily requirement.

Post-operative chest X-Ray is mandatory to exclude any pneumothorax or collapse Fig (5) .Antibiotics administered pre-operatively , continued post-operatively for five to seven days.

#### **RESULTS:**

Five of the patients were male constituting (62.5%), the rest were female constituting (37.5%). The youngest baby was only a twenty-five days old femalewho presented with repeated vomiting after feeding leading to recurrent chest infection. The oldest child was a six years old femalewho presented with severe recurrent chest infection & vomiting. The majority of patients(five cases) (62.5%) were seen within the 1<sup>st</sup>. year of life as shown in Fig (1).

Vomiting was the major symptom which was present in all the cases and it has been characterized as effortless regurgitation of undigested food occurring when the infant is recumbent .Recurrent aspiration and pneamonitis leading to recurrent chest infection was present in all the patients but with varying degrees of severity. Failure to gain weight and subnormal physical development were characteristic features .Anemia resulting from chronic blood loss secondary to peptic esophagitis was a common finding to all the patients but frank

haemoptysis leading to severe anemia was reported in only one (four years old ) male patient .Actually these symptoms were mostly seen in all patients but vomiting was more evident in older children .Table (1) summarized the cases , their ages , gender , clinical presentation & duration .

Plain chest X-Ray was done to all the patients and the major finding was the presence of an opacity with air fluid level in the right lower hemithorax. This opacity was obscuring the right hemi-diaphrgam and this was seen in seven patients Fig (2) and in only case there was absence of the gastric air bubbles in the left subdiaphragmatic region.

The h.h was cleanly shown in the barium study of all the patients as seen in fig (3) & (4).

laprotomy, all patients were found to have in the right chest cavity above the diaphragm, the left pillar of the diaphragm was well formed, the right pillar of the diaphragm was thin and laterally displaced, the esophageal hiatus was wide. There was a well formed sac formed from the posterior parietal peritoneum which lies in the extra-pleural space (i.e.) all had large hiatal orifice and the stomach was the main organ to be herniated. In five of the children the esophagogastric junction was displaced into he chest.

In only one patient the pre-operative findings of delayed gastric emptying on barium meal that an additional procedure of pyloro-myotomy was added .

All the cases had a smooth post-operative course with minimal complications *wound infection* was detected in only one patient and was treated properly by intra-venous antibiotics according to culture and sensitivity test.

Post operative *right lung collapse* was detected in one patient due to accumulated secretion and the already present chronic chest infection that necessitated bronchoscopy for clearance of secretion and allowing the lung to re-expand fully.

#### **DISCUSSION:**

A right intra- thoracic stomach secondary to a congenital hiatus hernia is a rare form of this congenital anomaly (R-4). It is characteristically situated at the right of the lower thoracic esophagus where part of the gastric fundus has herniated into the chest.

We report eight cases of this uncommon variety of the congenital hiatus hernia during a six years period and this rarity is comparable to studies done by Haddad & et all, Hashemi & et all and Arthur & et all (R 5, 6, 7)

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The etiology of the hiatus hernia and gastroesophageal reflux in infancy and childhood is not well understood. Wylie et all believed that congenital shortening of the esophagus produces hiatus hernia (R. 8) However at operation the fact that the herniated gastric cardia can be brought easily below the diaphragm argues against this concept as stated by Belsy R (R.9).

Congenital hiatus hernia is usually secondary to a developmental defect in the right crus of the diaphragm i.e. congenital anatomic defect , however The etiology of this type of hernia is still inadequately understood (R .10).

At the present time the etiology is still unknown and a single explanation for the incompetence of the gastro-esophageal junction in infancy and children remains unproved but as a consequence of this congenital enlargement of the esophageal hiatus there will be a progressive herniation of the abdominal content into the thoracic cavity and this herniation is facilitated by the negative intra-thoracic pressure generated during inspiration .If the herniation progresses, the entire fundus and proximal antrum may migrate into the thorax and at this stage organo-axiall volvulus inevitably occurs (R-11)

Most of our patients were males constituting (62.5%) of the cases as is the case with other studies and most of the patients were seen in early childhood Table (2) shows a comparison between three studies as regard the male predominance and the duration of the study.

Our cases presented with vomiting and recurrent chest infection and the younger the patient presentedmore with vomiting while the older presented more with chest infection so the symptomatology of our cases were non specific in the form of recurrent attacks of vomiting and/ or repeated attacks of chest infection .This coincides with the studies done by A H Al-Salem , M.woodward & Musafa imanoglu (R.12, 13, 14).

A definite radiological abnormality was seen in all the patients. either an opacity with air fluid level in the right lower hemi-thorax which obscures the right hemi-diaphragm as seen in seven of the cases and in only one patient there was absence of the gastric air bubbles in the left sub- diaphragmatic area and these findings were nearly the same as in other studies (R 10, 13, 14) .The diagnosis was confirmed by barium meal which was done to all the patients and it clearly demonstrated the exact position of the stomach in the right hemi-thorax and the position of the gastro-esophageal junction and for us it is the diagnostic modality of choice to

confirm this anomaly and this coincides with other studies (R.10, 13, 14 CT-Scan was not used in our study although it is regarded by some authors as the most important diagnostic modality, not only to establish the diagnosis but also demonstrated the exact content of the hernial sac and any associated lung abnormality (R .15) Rodrigo J described the pre-natal Recently. sonographic diagnosis of hiatus hernia in the fetus which can be done by ultra-sound at 33/52 week of gestation by detection of a hypoechoic mass in the posterior mediastinum in continuity with the intra-abdominal stomach bubbles .Post natal management was planned as an elective procedure (R-16). This facility is not available to us .

No pre-operative serious complication was encountered in our study in contrast to other studies done by Rawat J & Al-Salem AH (R 17 , 18), as they described cases of pre-operative gastric volvulus in an infant leading to an acute presentation.

All cases included in this study received a good pre-operative preparation as regard feeding in upright posture with frequent small feed and an antibiotics cover . Pre –operative esophagoscopy was not used in any patient, but bronchoscopy was done pre-operatively for two patients aiming for clearing of the accumulated secretion in the endo-bronchial tree of the already present chest infection and in one case post-operatively due to right lung collapsed .

Although in infants and children a successful repair of an uncomplicated hiatus hernia can be accomplished with equal ease through the chest or through the abdomen but we found that managing this type of hiatus hernia by laparatomy was the procedure of choice as the exposure was excellent ; infants and children tolerated the abdominal approach well, thus avoiding the morbidity of thoracotomy in the younger age group . It gives access to the pylorus if there is a concomitant outlet obstruction (As seen in one of our cases for whom pyloro-myotomy was performed) and the hernia is easily reduced and this also coincides with the studies done by Rawat J & Al-Salem AH (R 17.18).

The operative reflux and in this we agree with the studies done by M.Woodwark & Mustafa imanoglu (R 13, 14). Adding Anti – reflex fundiplication will prevent recurrence .In general, Nissen ' fundoplication is safe and effective with alow mortality rate (R.19) Immediate post-operative chest X-Ray is mandatory to exclude pnemothorax or lung collapse .

Feeding resumes from the third post-operative day .

All the patients rananuneventfull post-operative course apart from simple wound infection which was treated conservatively and bronchoscopy was needed in one patient to treat post-operative right lung collapse .

None of the patients required post-operative ventilatory support

Or post-operative tube thoracostomy

The reported self limiting complications of Nissen ' fundoplication such as gas bloat syndrome , dysphagia ,and Achalasia of the cardia

(R.20) were not encountered in our cases .

No post-operative recurrence of symptoms or the hiatus hernia during the follow up period was recorded .

#### **CONCLUSION:**

Congenital right intra-thoracic hiatus hernia presents a diagnostic challenge to the pediatrician, pediatric and thoracic surgeon.

-It should be included in the differential diagnosis in children with repeated attack of chest infection and vomiting or any infant or child with persistent respiratory and or gastro-intestinal symptoms

Laparotomy was the approach of choice in this type of congenital hiatus hernia and adding anti-reflux procedure was mandatory.

-Team work between the (pediatric and the thoracic) surgeons resulted in a successful and fruitfull outcome.

Early recognition of these uncommon cases is essential for early repair thus reducing the future morbidity and mortality .



Figure (1) :- Patients distribution as regard the age



Figure (2)- plain X-Ray Chest

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Fig (3) Barium Meal



Fig (4) :- Barium meal lateral view

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Fig (5) Normal post-operative Chest X-Ray

Case No	Sex	Age	Presentation		Duration
1	Female	3Y.	Recurrent chest infection	Vomiting	1 year
2	Female	25 Days	Vomiting		Since birth
3	Male	4 Y.	Haematemesis		1 month
4	Male	1 Y.	Vomiting		Since birth
5	Male	1 Y.	Recurrent vomiting	Cough	Since birth
6	Male	1 Y.	Recurrent vomiting	Cough	Since birth
7	Female	6 Y.	Recurrent chest infection		1 year
8	Male	11/12	Recurrent vomiting		Since birth

Table (1):Cases Resume

Name of the	author No.of cases	Male Fem	ale Dura	ation of the study
A H Al-Salem	6	4	2	1989-1997
Mustafa imanoglu	5	3	2	1992-2004
Waleed M. Hussain	8	6	2	2000-2005

 Table (2):Comparison between different studies

#### **REFERENCES:**

- 1. Hubertt BC. Familial right thoracic stomach .Pediatric 1987 ;79, 430-431.
- 2. William C., Michael D., Bork D etall Hiatus Hernia, The Fetus Net, 1994 ;06,02-01
- **3.** Gerson DE , Lewick .Am Intrathoracic stomach ,When does it obstruct ?Radiology 1996 ; 119 ,256-264 .
- **4.** Kim ME, Pyun HW, Kim MR, Kim By ,Lee JG, Right Intrathoracic Stomach Secondary to Congenital Hiatus Hernia with organo axial Torsion, J.Korean Radiol Soc2001; 45,79-82.
- 5. Haddad M.C. Youssef BA, Sammak bm, Duff A Rt.Intra-thoracic stomach secondary to congenital hiatus hernia and organoaxial torsion .AJR : 1996 ;167, 66-68.
- 6. Hashemi M , Sillin LE , Peter JH Current concept in the management of PEHH J , Clin Gastroenterol 1999 ;29, 8-13.
- 7. Arthur M , Olsen , Colin B , and et all Hiatus hernia in children Chest 1960 ;38, 495-505.
- 8. Wylie W.G , and Field E.C. The etiology of intermittent esophagitis , regurgitation and hematemesis in infancy .Arch . Dis .Child .1946 ; 21, 218.
- 9. Belsy R ; Surgery of the D(Children) Brown .J. M(ed) 1963, 762
- **10.** Jawad AJ ; Al samarraie Al ,Almofala S et allCongenital PEHH in infancy Pediatric Surgery International 1998 ; 13, 91-94.
- D.E Maziak , TR -j todd & F.G. Pearson Massive hiatus hernia Thorac Cardiovasc .Surg., 1998 ; 1/5 , 53-62.

- **12.** A H Al-Salem , Congenital PEHH in infancy and childhood .Saudi Med J . 2000 ;21, 164-167 .
- **13.** M. woodward , D.C.G., Crabbe , E.M.Kicly et all Congenital intra thoracic stomach .Pediatric Surgery International 1996 ;11 ,116-118 .
- **14.** Mustafa imanoglu , Ali Cay , Polat Kosccu & et all Congenital paraesophageal hiatal hernia ;pitfalls in the diagnosis and treatment , J.Pediatr.Surg. 2005 ;40, Issue 7 ,1128-1133.
- **15.** Raghunath B , Dipali Paras , R . KothariCongenital paraesophageal hernia presenting with severe GE reflux.Indian J Pediat .2007 ; 74, 310-311 .
- **16.** Rodrigo Ruano et all , Prenatal sonagraphic diagnosis of hiatal hernia ( Prenatal diagnosis J . Wiley interscience 2003; 24, issue (1) ,26-30 .
- **17.** Rawat J, Rasid KA, SINHA sk, Singhs Congenital PEHH with intrathoracic gastric volvulus in an infant .Pediatric Surg.Int. 2007.
- AL-Salem AH , Intra thoracic gastric volvulus in infancy .Pediatr Radiol 2002 ; 32, 614.
- **19.** R Minjarez , RC . Jobe BA Surgical Therapy to GERD GI Motility on line ,Doi ; 10 . 1038 , Gimo 56 .
- **20.** Waring JP Post fundoplication complications .Gastro enterol .Clin .Nort .Am 1999;28,1007-1019)