

were taking care of the child, once again did not agree for earlier surgery. This child also lacks mental and physical maturity of his age. Modified Heller's myotomy was done, by trans-abdominal approach, without any antireflux procedure. Our mainstay of diagnosis in both the cases was barium swallow since we do not have the facility for manometric studies and endoscopic examination.

Case No. 3:

This neonate, a full term female of 5.8 lbs, was born at home in Dera Ghazi Khan. The complaints were regurgitation after every feed, since birth, not relieved by antiemetics prescribed by Paediatricians. When we saw the patient she was severely dehydrated. Her hydration and electrolyte imbalance was corrected. Modified Heller's myotomy was done through trans-abdominal approach at the age of 6 weeks. The result of the operation was excellent and the child was discharged on the 9th post-operative day.

Case No. 4:

This child reported at the age of 3½ years with the complaints of regurgitation since birth after every feed, not relieved by antiemetics. He was diagnosed as a case of cardiac achalasia on barium swallow. Modified Heller's myotomy was done with trans-abdominal approach. Post-operative results were excellent.

Case No. 5:

A male infant, 3 months old, presented with complaints of regurgitation since birth after every feed, not relieved by antiemetics. Child was seriously malnourished and he was moderately dehydrated. It was diagnosed as a case of cardiac achalasia on barium swallow. Parents were advised surgery, which they refused.

TABLE 1

SYMPTOMS	Numbers	Percentage
Regurgitation :	5	100%
Repeated chest infection :	4	80%
Failure to thrive :	5	100%
Lack of mental maturity :	3	60%

TABLE 2

SEX RATIO

SEX	Number	Percentage
Male :	4	80%
Female :	1	20%

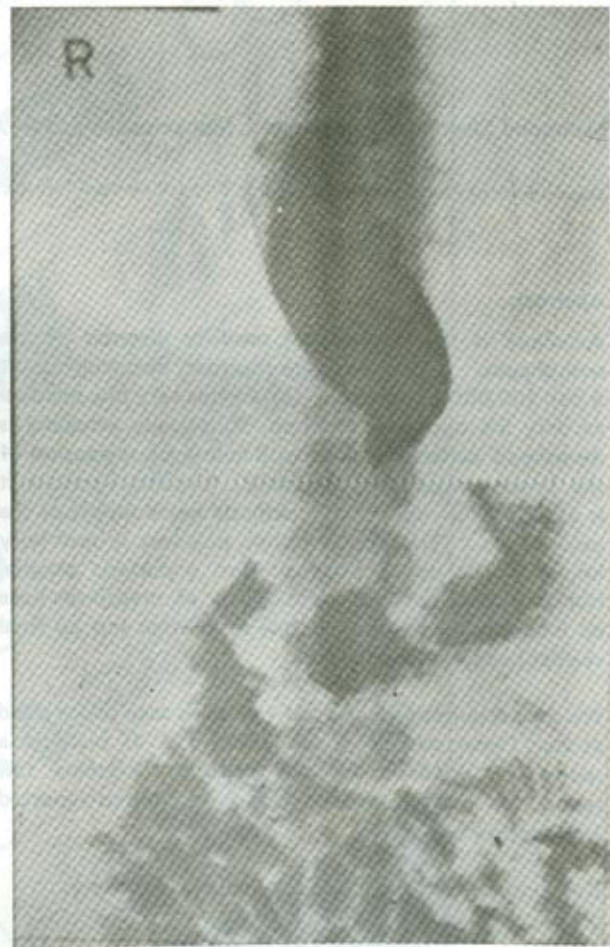


Fig. 1. Achalasia cardia.

Operative Procedures : Modified Heller's Myotomy was done in all patients without an anti-reflux procedure through the abdomen.

Mortality : No peri-operative mortality.

Follow up: Unfortunately, we do not have any facilities for endoscopy and manometry so only clinical follow up

Congenital Oesophageal Achalasia

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Abstract:

Achalasia is the commonest motility disorder of the oesophagus. It is a very rare disorder, occurring about 6 per 100,000 population. Out of this very low incidence, only 5% occurs in children. Classically, achalasia is disordered peristalsis along with failure of relaxation of the lower oesophageal sphincter. Broadly speaking, it is a disorder of the parasympathetic nerve supply of the oesophagus. The actual lesion however is not fully known. The dorsal vagal nucleus, myenteric ganglia, fibres in the wall and connections of myofibrils have all been implicated at one time or another but no final verdict has so far been given.

We are presenting a report of five cases seen and treated at our Department between 1980-87. It includes a report of achalasia in two brothers. Before this report, the total cases of achalasia in siblings reported in literature was 9.

Key words: Achalasia, Siblings, Heller's Myotomy.

Material and Methods:

All patients were admitted and treated at Mayo Hospital, Lahore.

Case No. 1:

Full term male having a weight of 6.5 lbs. was born at a private hospital in Lahore and was the 1st child of

parents. Patient started having regurgitation immediately after birth and was looked after by a Paediatrician, who prescribed all kinds of antiemetics but to no avail. Ultimately, the parents consulted us at the age of 4 weeks. Barium swallow was done, which showed dilatation of the esophagus with tapering at lower end. All the other systems were normal. The parents were advised operative treatment but the Paediatrician convinced them against surgery. Patient also used to take semisolid and solid diets through nasogastric feeding. When he reported again, his mental age was 2 years and his physical age was 2½ years. The patient had history of repeated chest infections.

He was operated in Jan. 1986 at the age of 4 years by trans-abdominal approach. Modified Heller's myotomy was done without any anti-reflux procedure. Post-operatively, the result was excellent. He was discharged on the 8th day after operation, from the hospital. He started taking all kinds of food including solids. He has started improving both physically and mentally.

Case No. 2:

This full term male of 6 lbs was also born in Lahore at a private maternity home fifteen months after the first one. He presented with the complaint of regurgitation since birth. He was diagnosed at the age of 2 weeks with barium swallow, which showed dilatation of esophagus with parrot's beak deformity at lower end. The severity of symptoms was less in this child. He initially had naso-gastric feeding but later on he could take liquids on his own, and the naso-gastric tube was removed. The child was operated upon at the age of 3 years in April 1986, since the Paediatricians who

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is available. All patients are well with the longest follow up of 3 years and shortest follow up of 6 months. One patient was lost to follow up.

Discussion:

Achalasia is a disease known to medical profession since 1674, described by Thomas Wills. He treated this condition with dilatation. Oesophagomyotomy was proposed by Gottstein in 1901 and was first performed by Heller in 1914. Heller performed myotomy both anteriorly and posteriorly. Presently modified, Heller's myotomy is performed which is only anterior myotomy and was introduced by Groeneveldt in 1918. Controversy still lingers on regarding the choice between the surgical treatment and forceful dilatation in adults, but there is no dispute in the treatment of children up to the age of 10 years. Oesophagomyotomy is the treatment of choice in this group. It is interesting to see that cardioplasty and oesophagostomy were the procedures which were being widely used in Great Britain, Germany and USA at the time when Heller was performing oesophagomyotomy. After 36 years of the start of Heller's procedure, Barret and Franklin of St. Thomas and Brompton Hospitals described the major complication of this procedure, namely, reflux esophagitis. Around 1950, the operation started getting universal popularity.

All of our patients had symptoms of regurgitation since birth and their diagnosis was confirmed by barium swallow, in the neonatal age in two patients. There are other reports of achalasia in the neonatal and infantile age group in the literature. This supports the role of genetic inheritance as the etiological factor of this disease.

We did Heller's myotomy by trans-abdominal approach as, in our opinion myotomy upto arch of aorta is not required, as considered necessary by some workers in which thoracic approach is essential. We extend myotomy upto 2 cm proximal to the beginning of hypertrophied muscles and distally 0.5 cm on to the stomach.

It is worth mentioning here that in achalasia there is only regurgitation and not vomiting, because in vomiting there is forceful expulsion of gastric contents,

which is absent in achalasia. Moreover this regurgitation is aggravated on lying down. Achalasia is the absence of parasympathetic innervation of lower part of oesophagus while the innervation above this segment of oesophagus and in the rest of the gastrointestinal tract is normal. This suggests the idea of skip lesion. This idea has already been condemned for Hirschsprung's disease; it is yet to be proved for achalasia. Vagus nerve supplies the gastrointestinal tract upto the mid-gut. The fact that bilateral truncal vagotomy does not lead to dilatation of a part of mid-gut makes the etiology of achalasia more obscure. While considering inheritance as an etiological factor, it must be considered that there are no reports of achalasia in monozygotic twins, but still it is the genetic inheritance which probably explains the etiology of congenital achalasia in siblings besides other factors, which need to be discovered to reach a logical conclusion.

Conclusion:

All neonates and infants presenting with regurgitation, repeated chest infections and failure to thrive should be investigated to rule out this very important condition. Surgery is the treatment of choice. In fact, it is the only standard, acceptable treatment in this age group. The results of surgery are excellent and very rewarding.

References:

1. Achalasia in siblings, Stoddard C.J. and Johnson A.G. Br. J. Surg. Vol 69 (1982) 84-85.
2. Westley C.R., Herlitz J.J. Goldmans et al Infantile achalasia inherited as an autosomal recessive disorder J. Paed. 1975, 87:243-6.
3. Kilpatrick Z.A. and Milles S.S., Achalasia in mother and daughter. Gastroenterology 1972, 62:1042-6.
4. Mackler D. and Schneider R. Achalasia in father and son. Dig Dis 1978, 23:1042-5.
5. Singh H., Setlin R.S., Gupta H.L. and Khetagrel S.K.: Cardiac achalasia in child hood. Postgrad Med. J. 45:329.
6. Rozyck D.L., Ruben, F.J., Repin I, et al. Autosomal recessive defcession with short stature. Vitiligo muscle wasting and achalasia arch otolaryngol 1971, 93:194-7.
7. Buich R.G., Sputz L. Br. J. Surg., 1985, 72:341-343.