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Retrospective analysis of paediatric achalasia in India: Single centre experience

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ABSTRACT

Background: Developing countries at tertiary referral centre. The aim of this study was to share our experience of paediatric achalasia in Indian scenario. **Materials and Methods:** This was a retrospective analysis of children <16 years, operated for achalasia at our centre, from December 1998 to December 2011. **Results:** Total 40 patients (mean age 39 ± 4.29 months), including 1 patient of megaesophagus were operated over 13 years of period; 17 patients (associated congenital H-type tracheoesophageal fistula in one patient, non- responders/ lost follow-up for minimum of 3 years in 16 patients) were excluded from the study. The response rate of parents in follow-up was 60.0%. Mean symptoms duration was 27.88 ± 2 months. Most common symptoms were regurgitation and failure to thrive (78.2%). Mean symptom scoring in follow-up after 3 year was 1 ± 0.7 compared to 5 ± 0.51 at the time of admission ($P < 0.012$). One infant expired (mediastenitis), one developed adhesive intestinal obstruction and one needed posterior re-myotomy (for megaesophagus). There were no treatment failures in mean follow-up of 40.2 ± 5.07 months. **Conclusions:** Cardiomyotomy with partial fundoplication is the best modality of treatment for paediatric achalasia cardia, even from parents' perspective.

Key words: Cardiomyotomy, fundoplication, pediatric achalasia cardia

INTRODUCTION

Achalasia is characterized by failure of co-ordinated relaxation of lower esophageal sphincter (LES) combined with ineffective peristalsis in body of the esophagus, resulting in functional obstruction of the esophagus.[1-5] Variants of achalasia (vigorous achalasia, short-segment achalasia) represents an early reversible stage, while failure to recognize wide spectrum symtomatology early in life, lead to progression of disease as end-stage esophageal dilatation (which may need esophagectomy).[1,5] Despite various advances in treatment for paediatric achalasia, the scenario in developing country is different because parent's perspectives are equally important in management of the disease. In this study, we shared our centre experience for achalasia in context of developing countries.

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MATERIALS AND METHODS

A retrospective record analysis of consecutive children with radiologically (oesophagogram) diagnosed achalasia of the oesophagus and had cardiomyotomy (Heller's operation) was carried out from December 1998 to December 2011, at the Department of paediatric surgery of the University hospital. Total 40 patients were operated over the 13 years- period. One patient was excluded from the study because of associated H-type congenital tracheoesophageal fistula and 16 patients, because no symptom scoring was obtained in follow-up for at least 3 years (lost on follow-up or non-responders). The demographic data, symptoms, their duration, diagnostic findings, surgical procedure details and complications were analysed.

Statistical analysis

Statistical data were analysed by SPSS version 17.0 for Windows. Continuous variables were expressed as mean values with two standard deviation and ranges (minimum to maximum), as well as in percentages. Statistical comparisons of pre-operative and post-operative variables were carried out with a non-parametric instrument (Mann-Whitney U-test). A linear regression model was used to evaluate the influence of different variables (symptom duration, degree of oesophageal dilation esophagogram) on the post-operative symptom scores. The P values of <0.05 were

considered significant for all purpose.

RESULTS

Over 13 years of period, total 40 children underwent surgery for achalasia cardia. Those belonging to rural areas were 82.5% (33/40). Fifty percent (20/40) parents' were educated. Sixteen patients were excluded because of lost on follow-up (10 cases) and no response to questionnaire send by mail (6 cases). The response rate of parents for follow-up questionnaire was only 60.0% (24/40). Final data analyses of 24 patients were done. Mean age at presentation was 60 ± 4.29 months (range 4 to 144 months). Male: female ratio was 32:8. The mean duration of symptoms was 27.88 ± 2.01 months (range 4 to 48 months). In none of our patients hereditary cause or syndrome (especially Allgrove syndrome) was noticed on clinical examination (hypoglycemia; hyperpigmentation of gums, tongue, lips, knuckles and pressure points; convulsions, coma; hyperkeratosis of palms/ or soles, fine palmer creases, incompletely developed dermatoglyphs; short stature; marked dental caries; microcephaly; hyperreflexia; ataxia/clumsiness, dysarthria; increased muscle tone, muscle weakness/ wasting; optic atrophy, sensory impairment, and mental retardation). So, further laboratory work up (Schirmer test and tear-film break-up time; adrenal function studies baseline as well as after adrenocorticotrophic stimulation test, and plasma rennin activity) for the Allgrove syndrome was also not done in any patient. The symptoms were mainly regurgitation 79.1% (19/24 patients) and failure to thrive 79.1% (19/24 patients), followed by recurrent pulmonary infections 73.9% (17/24 patients) [Table 1]. The pulmonary complications were ranged from hyper-reactive airways to pneumonia. The esophagogram showed typical 'bird's beak' appearance in 23 patients, while megaesophagus in one (4.3%) patient. The degree of esophagus dilatation had no correlation with the duration of symptoms (P value >0.05). The diagnostic esophagoscopy in five patients showed dilated esophagus with difficulty in negotiating the scope through the cardia and stages I to II oesophagitis (Savary and Miller staging). All these five patients received antacid therapy in preoperative period to 6 months in follow-up.

Table 1: Symptomatology

Symptoms	Number of patients	Percentage
Dysphagia	15/24	62.5
Chocking	12/24	50.0
Cough	10/24	41.6
Regurgitation	19/24	78.2
Weight loss	19/24	78.2
Recurrent chest infection	17/24	73.9
Retrosternal pain	12/24	50.0

The oesophagus was approached through laparotomy in 16, laparoscopy in seven, and left posterolateral thoracotomy in one patient. In 95.8% (23/24) patients, fundoplication (5 Nissans', 14 Dor, 1 Toupet and 3 Boix-Ochoa) performed with myotomy, while one patient, who operated through thoracotomy, needed it later. Intraoperatively, 20.8% (5/24) patients had mucosal perforation (four during laparotomy and one during laparoscopy). The site of perforation was oesophagus in one and upper stomach in four. The perforations were repaired and strengthen by Dor fundoplication. In two patients diversion gastrostomy (>1 cm diameter mucosal perforation) was additionally performed, while in rest three patients, nasogastric tube was left for one week. In both children, the gastrostomy was closed after four months of surgery.

In the immediate post-operative period, three patients developed bronchospasm, which was managed conservatively. One infant expired in early post-operative period due to gastric juice induced mediasenitis. In six patients, who had no pulmonary or other complications, the mean hospital stay was 5 ± 0.43 (3-10 days). In rest 17 patients, the mean hospital stay was 15 ± 0.48 (13 to 20 days). In the mean follow-up of 40.2 ± 5.07 (12 to 100 months), all patients gained adequate weight and were relived of regurgitation and recurrent chest infections. Mean symptom scoring in follow-up after 3 years was 1 ± 0.70 compared to 5 ± 0.51 at the time of admission ($P < 0.012$). The symptom duration had no relation with the preoperative or postoperative symptom scoring ($P > 0.05$). Post-operative esophagogram done after 10 days of surgery showed disappearance of bird's beak in all patients. The patient having megaesophagus needed posterior re-myotomy after 1 year because of non-resolution of symptoms, but did well after re-myotomy in 1 year of follow-up. We have not found recurrence of disease, dysphagia (obstruction) due to fundoplication or peptic stricture due to acid reflux in any patient. One patient with gastrostomy presented as adhesive intestinal obstruction three months after surgery, which was managed conservatively.

DISCUSSION

The incidence of achalasia cardia in children is 0.4 to 1.1 in 100,000 and prevalence is 7.9 to 12.6 per 100,000.[1-4] Fewer than 5% of patients having symptoms of achalasia cardia are younger than 15 years of age.[2-4] Ellis, was the pioneer of the transthoracic

approach, while Ernest Heller (1913) first successfully performed cardiomyotomy.[5,6] Since than many studies was performed on paediatric achalasia and various guidelines has been established for it.[3,7-13] Familial achalasia is present in less than 1% of all cases.[1,14] Few cases of Allgrove syndrome or triple A syndrome were reported in the children.[14] But, we did not found either of these in our study group. We had the mean age of presentation slightly higher than Pastor *et al*. series, but much lower than Lee and Askegard-Giesmann *et al*. series.[2,7,13]

Typically, adults and older children have progressive dysphagia to solids and liquids, chest pain and weight loss.[1-6] But, symptoms in infants and toddler are chocking, cough, recurrent pneumonia and failure to thrive.[1-6] The vomitus has non-clotted milk in breast feeders, while the children vomits food eaten several hours ago.[2,13,15,16] Majority of our patients were toddlers, so regurgitation, failure to thrive, and pulmonary complications were the predominant symptoms rather than dysphagia.[2,13] The pulmonary complications ranged from bronchospasm/asthma (due to recurrent aspiration), stridor (tracheal compression from dilated esophagus) to 'Bull frog neck' appearance (due to compression tracheal above the larynx) in its most severe form.[15-18] Because the pulmonary complications and malnutrition/ failure to thrive significantly increase the morbidity and mortality of child, it is extremely important for clinicians to be aware of the diverse symptomatology in children. Most of these patients turned up in paediatrician clinics rather than to the paediatric surgeons, hence the diagnostic delays in paediatric achalasia are usually due to misinterpretation of these typical findings.[18-20] We had higher pulmonary symptoms (73.9%) than Eckards' series (33%).[10] The probable cause of higher pulmonary infection was faulty feeding practices by illiterate parents in the developing countries.[20]

The manometery remains the diagnostic modality of choice for achalasia (highest sensitivity), but we lack the facility even at tertiary referral centre, because of inequality of health resources in India.[21-23] If we further refer these patients to higher centres (few in developing countries), where these facilities are available, a long waiting list at these centres, assessability and affordability of parents' further delays the treatment.[21-24] Beside these reasons, as was already reported in the literature that resting LES pressure level has no effect on the outcome of surgery.[5] Hence, assuming no other cause for bird's beak sign on esophagogram, we used it as the sole diagnostic modality.[1-6] In the absence of manometery we have misdiagnosed one case of congenital H-type trachea-esophageal fistula as achalasia cardia. In this patient final diagnosis was made during surgery, when following cardiomyotomy exposed mucosa showed excessive mobility with each act of ventilation made by anaesthetist. In these cases also, cardiomyotomy with excision and ligation of fistula is recommended.[24] S0, we had managed the case by bronchoscopic guided catheterization of fistula with ureteric catheter and through neck exploration excision and ligation of fistula.

There were controversies whether degree of esophageal dilatation on esophagogram depends on duration of symptoms or not? Some authors believe that it takes 16 to 21 years for the development of megaesophagus, but the author did not find any co-relation between both as none of our patient (even megaesophagus) had such prolonged course of illness.[17,25,26]

We have used enteral nutrition via nasogastric tube in pre-operative period to all those patients who presented with failure to thrive and via gastrostomy tube in postoperative period in whom oral feeds deferred (large mucosal perforation). The enteral nutrition via nasogastric/gastrostomy provides positive nitrogen balance and further decreases the risk of pulmonary aspiration in these patients.[27]

In developing countries parents' affordability, availability of facilities, local expertise, long-term results of different modalities (pharmacological/dilatation/botox injection), parents preferences, compliance with treatment and follow-up is the major detrimental factors for the treatment of achalasia cardia, rather than patients' characteristics.[1,13,21-23] Other relevant issue from parents' perspective is poor economic status (expense on repeated transportation to visit tertiary centre), illiteracy, lack of compliance with repeated dilatation or botox injections and follow-up fundoplication.[15,16] In the background of above-mentioned factors, we had not used pharmacological treatment (temporary effect), botulinum toxin, pneumatic dilatation (repetition rate 17%).[2,4,5,17,19] Further, pneumatic dilatation is not preferred modality of treatment if pulmonary pathology is present, and in our study group majority suffered from it.[2] Because the long-term permanent treatment success rate is highest with myotomy (47% to 82%), hence, we used surgical treatment as the sole mode of treatment in all our patients.[2,4,17-19]

The laparoscopic Heller's cardiomyotomy evolved as the procedure of choice in paediatric as well as

adult achalasia cardia (less need for redo-myotomy or esophagectomy).[28,29] The author recommend pharmacological treatment only for patients having high risk for invasive procedures or awaiting definite treatment.[2,4,5] We have used extended cardiomyotomy, because author believes that longer myotomy results in an even better and sustained resolution of dysphagia as compared with the traditional cardiomyotomy (extending 1 to 1.5 cm onto the gastric wall) and to avoid inadequate myotomy (the most frequent reason for 'early' reoperation).[1] The underlying mucosa was mobilized for one third to half of its circumference to prevent early fusion of the myotomy, which is the most common cause of persistent or early recurrent dysphagia.[1] Contrary to Ali Avano~lu *et al*. opinion that in children anti-reflux treatment should be added only if incorrect myotomy (extra large) is done, we added anti-reflux procedures as prophylactic measure in all as previously proved that long-term poor surgical results of cardiomyotomy (92%) are due to late complications of severe GERD (peptic ulceration subsequent stricture and malignancy in Barret oesophagus) and not from incomplete myotomy.[1,2,4,29,30] We preferred Dor procedure because it is equally effective to Nissens' but has lesser rate of dysphagia (2.8% vs 15% in Nissens' fundoplication).[31]

Although we had mucosal perforation in five patients as compared to none in Lee *et al*., and 12% in Boeckxstaens study, but we had no recurrence of symptoms, or mortality (except one) in follow-up as compared to 40% in Lee series and 0% to 14% in other series.[1,13,32,33] We added gastrostomy in which there was large mucosal perforation. Gastrostomy, besides helping in healing of perforation by diverting acidic juices, also decreases the risk of aspiration pneumonia in post-operative period because of acquired oesophageal dysmotality in achalasia.[22,34,35] We had used chest infection instead of chest pain in scoring (contrary to Eckerd's scoring) because latter symptom is difficult to interpret in the children.[16,18,19] The scoring used by us was semi-quantitative and can be obtained from parents' by simple questionnaire by mail also. Author believes that the semi-quantitative measures can make various studies comparable.

There was no case of malignancy in this series, contrary to 1% to 33% reported in different series, probably our period of follow-up was too short to comment on it.[36,37]

To conclude, our study is the largest series of pediatric achalasia ever described in developing countries. Clinicians should identify the wide spectrum of symptoms of paediatric achalasia for early treatment. Enteral feeding should be used whenever possible, especially who fails to thrive. The gastrostomy is the beneficial additional procedure in patients of larger mucosal perforation. Despite of advances in management of achalasia in developing countries, still the best treatment is cardiomyotomy with fundoplication both from parents as well as surgeons perspective.

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