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ACHALASIA CARDIA IN INDIAN CHILDREN- EXPERIENCE FROM A TERTIARY CARE INSTITUTE IN CHENNAI, INDIA

Gastroenterology

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ABSTRACT

Introduction: Achalasia cardia a rare oesophageal motility disorder is not uncommon in children.
Aim and objectives: To describe the clinical profile of achalasia from a single pediatric tertiary care centre.
Materials and Methods: Retrospective analysis of case records over a period of 10 years from March 2008 to March 2018 at institute of child health and hospital for children, Chennai, India.
Results: 26 children had achalasia and 11 (42.3%) were males. The mean age was 42.08±36.33 months and 19 (73.07%) were less than five years at diagnosis. Vomiting was the most common symptom (88.4%) followed by failure to thrive (80.77%). Six had features of Allgrove syndrome. Heller's cardiomyotomy with or without fundoplication was done in 24 (92.3%) and pneumatic dilatation in two.
Conclusion: Achalasia cardia is not uncommon and should be considered in children presenting with vomiting, dysphagia and failure to thrive. Surgery remains to be safe and effective.

KEYWORDS

Achalasia, Children, Manometry, Heller's Myotomy, Pneumatic dilatation

INTRODUCTION:

Achalasia is a rare oesophageal motility disorder characterised by absence of oesophageal peristalsis and failure of relaxation of lower oesophageal spinchter (LES) on swallowing[1] with an estimated incidence of 0.11/1,00,000 in children, in contrast to adults with an incidence of 1/1,00,000.[2] Childhood, achalasia account for only 5 % of the total cases and the common symptoms include vomiting, dysphagia and weight loss similar to gastro oesophageal reflux disease.[3] We herein describe the clinical profile of achalasia in children.

AIM AND OBJECTIVES:

To describe the clinical profile of achalasia in paediatric age group.

MATERIALS AND METHODS:

Retrospective analysis of Children with achalasia was done by reviewing the medical records database of Institute of child health and hospital for children, Chennai, India for a period of 10 years (March 2008-March 2018) with respect to demography, clinical features, diagnostic work up and treatment.

RESULTS:

Demographics:

Total number of children with achalasia was 26 and 11 (42.3%) were males. The mean age was 42.08±36.33 months and median age was 36 months at diagnosis. Age range in the cohort was between 2 months to 120 months. Mean age was 48.73±41.32 months and 37.2±32.82 months in boys and girls respectively without statistical significance p=0.44.

30.7% children (8/26) belonged to urban region and consanguineous parents in six (23.07%) with family history of achalasia in a 8 year old female child, born of 3rd degree consanguineous marriage. Table 1 illustrates the demographic details.

Table 1: Demographic data

No of patients	26
Male: female	11:15
Rural:Urban	18:8
Consanguinity	6(23.07%)
Family history	1(3.84%)
Mean age±SD at diagnosis	42.08±36.33 months

Clinical features:

Median duration of symptoms was 6 months (1-72 months). Clinical symptom observed were vomiting, failure to thrive, dysphagia to both solids and liquids, regurgitation of feeds, recurrent pneumonia due to aspiration and cough. Six children had features of Allgrove syndrome (alacrimia, achalasia and addisons). without any statistical significance in the symptomatology of achalasia between Allgrove syndrome group (n=6) and isolated achalasia group (n=20). Six children had other associated features like mental retardation(3/26), bronchiectasis(1/26), ectopic kidney(1/26) and blonde hair(1/26).

Table 2 gives the clinical details of our study. Comparison features of clinical data based on age at diagnosis and rural/urban background given in table 3 and 4 respectively.

Table 2: Clinical profile

Clinical features	n (%)
Vomiting	23(88.4%)
Poor weight gain/weight loss	21(80.77%)
Dysphagia	11(42.3%)
Regurgitation	10(38.46%)
Recurrent pneumonia	8(30.77%)
Cough	5(19.23%)

Table 3: Comparison of clinical data based on age at presentation

Clinical features	< 5 years age at diagnosis (n=19)	>5 years age at diagnosis (n=7)	P value
Vomiting	16(84.21%)	7(100%)	0.54
Poor weight gain/weight loss	17(89.47%)	4(57.14%)	0.10
Dysphagia	4(21.05%)	7(100%)	0.0005 *
Regurgitation	9(47.37%)	1(14.29%)	0.19
Recurrent pneumonia	5(26.32%)	3(42.85%)	0.64
Cough	3(15.79%)	2(28.57%)	0.59

*(*Data analysis showed statistical significance in dysphagia between < 5 years and > 5 years age group.)*

Table 4: Comparison of clinical data based on urban/ rural background

Clinical features	Rural (n=18)	Urban (n=8)	P value
Vomiting	17(94.44%)	6(75%)	0.22
Poor weight gain/weight loss	15(83.33%)	6(75%)	0.63

Dysphagia	9(50%)	2(25%)	0.39
Regurgitation	7(38.89%)	3(37.5%)	1
Recurrent pneumonia	6(33.34%)	2(25%)	1
Cough	5(27.78%)	0	0.15

(Analysis of data based on urban/rural background did not show statistical significance for any of the symptoms.)

Diagnostic workup:

Mean haemoglobin level was 10.95±1.29g/dl. Serum sodium and potassium levels were normal in all children. Barium swallow showing dilated oesophagus was present in all children. Upper GI endoscopy was done in all children and majority had dilated oesophagus with resistance at the LES (n=23/26;88.46%) and three (11.54%) had resistance. at LES without dilated oesophagus. Food stasis was present in 7 children (26.92%). Characteristic features of achalasia like absence of oesophageal peristalsis on swallowing, incomplete/absent relaxation of LES and high Integrated Relaxation Pressure (IRP) on esophageal manometry was present in all.

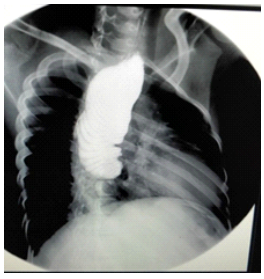

Figure 1: Barium Swallow

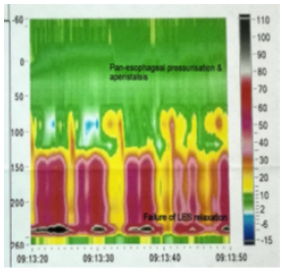

Figure 2: Oesophageal Manometry

Treatment:

24 children (92.3%) underwent Heller's cardiomyotomy with or without fundoplication, out of which a nine month female child with Allgrove syndrome had undergone surgery after failed endotherapy and two were treated with pneumatic dilatation alone. Treatment details are given below in table 5.

Table 5: Treatment details

Procedure	Number of children	Percentage
Laproscopic HM	1	3.84%
Abdominal /transthoracic HM	10	38.4%
HM+ Thal fundoplication	6	23.07%
HM+Dor fundoplication	6	23.07%
Pneumatic dilatation alone	2	7.70%
Pneumatic dilatation+HM	1	3.84%

Follow up:

Follow up duration ranged from 3 months to 9 years with mean of 3.3±2.60 years. Of the 24 children who underwent surgery, four children lost to follow up and outcomes were available on 20 children. Majority of the children (n=14;70%) are doing well. Two children died, one a 4 month old male child with blonde hair 3 months after surgery due to sepsis and other a 6 year old child born as a preterm/IUGR in the immediate post operative period after re-do surgery due to sepsis. One with Allgrove syndrome had hydronephrosis and urinary tract infection. Two children (4 years old male and 8 years old female) developed reflux esophagitis 9 years and 4 years after surgery and was managed conservatively. 5 year old male child with Allgrove syndrome developed stricture after surgery and was managed with endotherapy and currently under follow up. Of the children managed with pneumatic dilatation symptoms improved after repeated dilatations (3-5 dilatations) except for one child who had undergone surgery after failed dilatation.

DISCUSSION:

This study describes, 10 year single tertiary centre experience regarding the demographic, clinical, management and follow up of 26 children with age ranging from 2 months -10 years and also first in the literature to describe the profile of achalasia in children predominantly aged less than five years, The mean age observed was 42.08±36.33 months, majority were (19/26(73.07%) under five years of age. This is in contrast to the studies done by Hussain et al,[4] Pastor et al,[5] Lee et al,[6] Singh et al[7] wherein the mean ages of children were 9.3 ± 4.7 years, 10.6 ± 4.6 years, 13 ± 6 years, 60 ± 4.29 months respectively. Though delay in diagnosis is known to occur even in adults due to misinterpretation of typical findings ,[8] our study had shown the time interval between the onset of symptoms to diagnosis was 6 months, similar to other paediatric studies. The most common symptomatology in our series was vomiting (88.4%), followed by poor weight gain/weight loss (80.77%) and dysphagia only in 42.3% in contrast to other studies done by Franklin et al (dysphagia-83%),[9] Lee et al (dysphagia-79%),[6] Hussain et al(dysphagia-81.8%),[4] Hallal et al (dysphagia-69.2%).[10] Probably this difference may be because, majority of the children were under five years in our study and dysphagia is often a predominant symptom in older children. Familial cases of achalasia reported in literature might suggest an inherited disease[11] but degeneration/loss of inhibitory neurons of myentric plexus of esophagus secondary to infection (Trypnosama Cruzi, viral infection[12]), circulating autoantibodies is the basic pathophysiology.[13] Association of achalasia with isolated glucocorticoid deficiency and alacrimia was first described by Allgrove and colleagues.[14] In our series six had Allgrove syndrome. Children with Allgrove syndrome have also associated neurologic abnormalities and autonomic dysfunction.[15] Mental retardation was present in 2 children with allgrove syndrome in our study and similar to a case reported in literature, one with Allgrove had renal ectopia.[16]

A combination of timed barium swallow oesophagogram,[17] upper GI endoscopy and manometry (gold standard)[18,19] which is helpful in diagnosis was performed. in all our patients. Achalasia can be classified into three subtypes by high resolution manometry based on the pattern of aperistalsis.[20]

Calcium channel blockers has limited role in children and partial response to nifedipine was reported in a study of four children by Maksimak et al[21]. Botulinium toxin injection has relapse rate of 50%.[22] Efficacy of pneumatic dilatation (PD) in children is variable ranging from10 to 90 % and response rates are less in younger age group.[23] Success rate also depends on the type of achalasia based on manometry with type 2 having good results.[24] In our study 2 treated with multiple dilatation had clinical remission and one had to undergo surgery after a failed endotherapy. Efficacy of dilatation done in our study was 66.67% similar to Pastor et al[5] wherein success rate was 70%. Majority had Heller's cardiomyotomy (HM) and it was found to be safe and effective similar to literature.[25] The most recent guidelines for the treatment of achalasia recommends myotomy with an antireflux procedure.[26] Success rate of HM varies between 60 to 95% and our study for HM was 75% similar to 70% by Hussain et al.[3] Oesophagectomy is a last resort in patients who are unresponsive to PD or HM therapy.[27] Latest therapy in the management of achalasia is Peroral Esophageal Myotomy (POEM).[28]

The limitation of the study is its retrospective nature. Though there are several studies on childhood achalasia with mean age of 8-12 years our study is unique of its kind because majority of children (73.07%) in our study are under five years age. Vomiting and failure to gain weight were the two most common symptoms in children under 5 years of age in contrast to dysphagia which was a predominant symptom in children above 5 years. Success rate for dilatation was better in older children and surgery was effective in the majority.

DECLARATIONS

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