

## TUBULAR DUPLICATION OF ESOPHAGUS: Rare congenital malformations require individualized and innovative procedures

Naeem-uz-Zafar Khan<sup>1</sup>, Col. Mansoor<sup>2</sup>, Anwar-ul-Haq<sup>3</sup>, Abid Qazi<sup>4</sup>

### ABSTRACT

Rare congenital problems generally have no standardized diagnostic or operative technique. Because of some variation within the spectrum, diagnosis and management has to be individualized and tailored according to the local findings to relieve the symptoms. We report a diagnostic dilemma and surgical management of a child with proximal tubular duplication of esophagus with complete remission of symptoms after surgery. To diagnose rare malformations a high index of suspicion is required when differential diagnoses are being considered.

**KEY WORDS:** Tubular Esophageal Duplication, Rare Anomaly, Individualized Treatment.

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### INTRODUCTION

Tubular duplication is a rare occurrence and is much less common than cystic duplication of foregut.<sup>1</sup> Epithelial lining is derived from primitive foregut diverticulum where as muscular elements are from splanchnic mesoderm. Failure of vacuolization of primitive septum has been considered a cause of tubular type of foregut duplication.<sup>2-5</sup> Clinical presentation varies from dysphagia, respiratory distress and occasionally gastro intestinal bleeding.<sup>6</sup> Generally duplications come to notice in older children and occasionally in adults.<sup>7-9</sup> Some times

diagnosis may be incidental on an X-Ray chest. Various diagnostic modalities include plain x-ray chest, C.T, M.R.I, Contrast Fluoroscopy and endoscopy.<sup>8-12</sup> Different investigations may add to additional information. Treatment may also have to be individualized according to presentation of symptoms, type and site of malformation. We are presenting a case of tubular esophageal duplication as an example to highlight various aspects.

### CASE REPORT

A six year old boy presented with four years history of difficulty in deglutition and vomiting especially to solids. He carried with him several x-rays of contrast barium swallow. These had been repeated almost every year for the last four years. On the basis of these he was diagnosed a case of proximal esophageal stricture and several attempts at dilatation had failed in other hospitals.

On presentation in x-rays there were three diverticular lesions the two in the cervical area whereas there was a larger one extended into the upper thorax and this looked exactly like that of upper pouch of a case of esophageal atresia. There was also an adjacent normal looking esophagus. (Fig-1) Because of the site,

1. Prof. Naeem-uz-Zafar Khan  
Department of Paediatric Surgery  
K.R.L Hospital,  
Islamabad.
2. Dr. Col. Mansoor
3. Dr. Abid Qazi  
Department of Children Hospital PIMS,  
Islamabad.
4. Dr. Anwar-ul-Haq  
Leeds General Infirmary U.K.

Correspondence:

Prof. M. Naeem-uz-Zafar Khan,  
House No.1, Street # 63,  
Sector F-7/3,  
Islamabad - Pakistan.  
Email: prnaeem@comsats.net.pk

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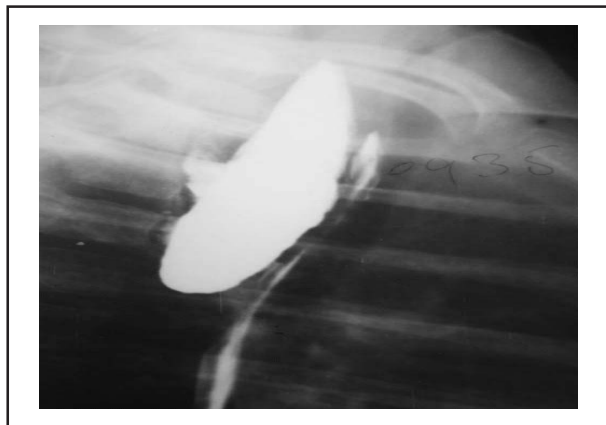


Fig-1: Tubular duplication of upper esophagus one blind ending. The other complete esophagus Dysphagia was his symptom.

we considered a provisional diagnosis of pulsion diverticulae or possibly proximal esophageal duplication. MRI was also not conclusive in diagnosing the condition. Endoscopy under anesthesia revealed two well formed lumens of the upper esophagus one ending blindly, while the other allowed the endoscope to pass in its entirety. Through fourth intercostals space and extra plural approach the site of duplication was reached. The postero-lateral blind ending duplicated tubular esophagus was isolated. Its medial surface was plastered to the normal esophagus. Both esophagi were vertically opened to enable side to side anastomosis. Thus the blind ending esophagus was incorporated to normal esophagus (Fig-2). Following this procedure recovery was uneventful and patient could take both liquids and solids from seventh post operative day. He has remained well ever since.

## DISCUSSION

Foregut duplications most frequently are found in the region of posterior mediastinum. Esophageal duplications are estimated to be 10 to 20 percent of these anomalies.<sup>13,14</sup> Tubular duplications as opposed to cystic esophageal duplications usually communicate to the normal esophagus. These are also much less frequent. Historically a tubular duplication was first described by Blasius in 1674.<sup>15</sup> the second case seems to have been spotted in 1907 by Kathe.<sup>16</sup> Most reported tubular duplications



Fig-2: Post operative appearance after incorporating the two tubes. While the other allowed the endoscope to pass in its entirety.

seem to present in late childhood or in adults. This will indicate their relative asymptomatic nature. This was also the case in our patient, who should have been diagnosed at the age of four years when he had first barium swallow performed. But at that time and in subsequent three years esophageal stricture was erroneously considered for his obstructive symptoms. A similar case like our patient was reported by Frank and Paul.<sup>17,18</sup> having two unequal parallel channels. This was noted on radiological examination in a five year old boy, who had presented with dysphagia. But he had communication with normal esophagus and only required symptomatic treatment and was well up to 17 years of age. At autopsy a 39 years old man with esophageal carcinoma was noted to have a duplicated esophagus. Because of asymptomatic nature of this abnormality possibility of greater frequency than it is clinically apparent has been postulated.<sup>19,20</sup> Respiratory symptoms may constitute dyspnoea, wheeze, dry cough, pneumonia nocturnal aspiration and gastrointestinal symptoms like regurgitation, dysphagia, vomiting failure to thrive, anorexia, epigastric pain, gastrointestinal hemorrhage and pyrosis. These symptoms may arouse possibility of existing foregut duplication. Proximal esophageal diverticulae seem to have the same embryologic origin as of duplication. In our case also this possibility was considered. Treatment of this condition must be individualized accord-

ing to the presenting symptoms and type of lesion. Various procedures that have been reported range from excision, enucleation, marsupialization, and internal drainage. Cauterization of mucosa, needle aspiration, and division of septum has also been performed.<sup>21</sup> in our case the blind ending tubular esophagus was incorporated with the normal esophagus by doing side to side anastomosis of two structures, thus converting these into one lumen

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