Congenital pouch colon: Present lacunae

The current issue of the JIAPS includes many articles on the rare variant of anorectal malformation - pouch colon. The condition of congenital pouch colon has been seen as a curiosity ever since its first description in 1912 by Spriggs in a London Hospital Museum specimen with absence of the left half of the colon and rectum. Various terms have been used to describe this entity including pouch-like dilatation of shortened colon, extrophia splanchnica, absence of colon and rectum, cystic dilatation of colon, short colon, colonic reservoir, congenital atresia of anus with short colon malformation, pouch colon syndrome, association of imperforate anus with short colon (AIASC) and congenital pouch colon. The term has now been included as rare variant, in the Krickenbeck classification of anorectal malformations.

Congenital pouch colon is a condition associated with anorectal agenesis, particularly seen in Asia, and is defined as an anomaly in which whole or part of the colon is replaced by a pouch-like dilatation, which communicates distally with the urogenital tract by a large fistula. The mesentery of this pouch of variable size (5-15 cm in diameter) is short and poorly developed, wall is very thick, the taenia coli are absent or ill defined, haustration and the appendices epiploicae are absent. The blood supply to the pouch is abnormal. The main pouch is supplied by the branches arising from the superior mesenteric artery that form a leash of vessels around it. There is no transitional zone between the pouch colon and the normal bowel. The pattern changes suddenly and sharply. The colon wall is thick and muscular with hypertrophied mucosa. The histology is also different and marked with two striking features; a very thick sub-mucosal layer and a criss-cross pattern of decussating fibers in the muscle coat. The normal longitudinal and circular pattern is found lost.

This condition is seen much more frequently in the northern, north western and central part of India and neighboring nations like Pakistan, Bangladesh and Nepal. Most of the patients have come from the states of Punjab, Uttar Pradesh and Delhi. Only a few reports have originated from China, Japan, Sweden, United Kingdom and other parts of the world. The cause of this unique geographical distribution has not yet been ascertained.

There is a need for following a uniform definition, classification and management policy.

1. The term congenital pouch colon is more descriptive and the infrequently used term short colon should preferably be avoided to avoid confusion in terminology.

2. For classification, it is much simpler and appropriate to use only the terms “Incomplete” and “Complete” pouch colon. It is incomplete congenital pouch colon if it involves only the terminal colon, where the length of the remaining normal colon is adequate enough for performing the pull through, without the need for doing a coloplasty, and complete congenital pouch colon is the one in which most of the colon is involved in the pouch formation, leaving insufficient normal colon for performing the pull through without the need for a coloplasty. The overall incidence of pouch colon is about 7-9% of all cases of the anorectal malformations in North India. Interestingly, during the past 25 years, whereas the incidence of complete pouch colon has decreased, that of incomplete congenital pouch colon has increased. Presently, incomplete pouch colon is almost three times more common than the complete variety.

3. As the dilated pouch is always abnormal in anatomy and function, surgical procedures should ideally include an excision of the dilated pouch with an end colostomy at birth and a definitive pull through later. A single stage pull through can be undertaken in the newborn stage in the incomplete type of the pouch colon, only if the condition of the baby permits. However, the problem is much more in patients with complete congenital pouch colon, as there is either no or little normal colon left which is not sufficient for performing the pull through. In this situation, a coloplasty procedure would be required to retain about 15 cm length of pouch colon in the form of a tube, to be brought out as an end colostomy. A window colostomy from the body of the pouch, though very easy and quick to perform in emergency, should be discouraged as it results with frequent prolapses, stasis and fecoloma formations. Also, a pull through procedure at the time of performing coloplasty should not be preferred in the newborn stage as it is associated with high morbidity and mortality. Any attempt to make a coloplasty longer than 15 cm, not only results in stasis and dilation of the segment but also causes frequent complications in the post pull through period.

Though there is enough literature on the descriptive and the management aspects of the condition from India, there
is a need for identifying the etiological factors as the condition is much more prevalent in a particular geographical area. There is also a need to study the vascular pattern and the motility abnormalities associated with the dilated pouch. Also, the experimental studies using various pharmaceutical agents on the muscle and the mucosal function of the pouch colon may address the problems in future commonly encountered with the use of coloplasty in these patients.

The present issue of JIAPS has been long awaited and includes contributed articles on pouch colon from different centers in India, addressing some of these issues.

REFERENCES


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