Jejunoileal Atresia in Association with a Multiple Gastrointestinal (GL-) Fistulae

David nii-Amon-Kotei, *Dip. med, Dr. med,* FACh, FAch, paed, FWACS
Department of Surgery
School of Medical Sciences
University of Science and Technology
Kumasi, Ghana

ABSTRACT

A long segment jejunoileal atresia in association with multiple internal GI-fistulas, and mimicking Hirschsprung's Disease in a six months old male infant with failure to thrive since birth is reported.

In analogy to a liquid reticulation system, and utilizing principles of fluid mechanics, the cause of the atresia and the clinical features of the case presented here are attributed to altered channeling and the rate as well as direction of flow of the intestinal contents. The usually mono-channeled GI-tract is converted into a multi-channeled system by the multiple fistulas through a parallel diversion. Complications and therapeutic uses of parallel diversions of GI-tract diversions have also been discussed.

KEYWORDS: Atresia, Hirschsprung's Disease, malabsorption, short gut, internal fistula, parallel diversion

INTRODUCTION

Theories advanced to elucidate the causes of intestinal atresias are not only many [2] but to some extent very divergent and are usually based on other primary pathological changes such as intussusception (a) and adhesions, strangulations and vascular damage following meconium peritonitis (b); the atresia therefore being secondary. This paper while presenting the association of a long segment jejunoileal atresia with multiple internal GI-fistulas also aims at contributing to the theoretical explanation of the etiological factors leading to atresias by using principles of fluid mechanics.

CLINICAL OBSERVATIONS

A six months old infant weighing 2.2 kg presented with a non tender distension of the abdomen and a chronic constipation which was interrupted occasionally by bouts of diarrhoea since birth.

On admission to the Komfo Anokye Teaching Hospital Kumasi-Ghana the patient was dehydrated, anaemic and in poor general condition. There were no other relevant physical or laboratory findings except the barium enema X-ray film which showed a grossly dilated left large intestine, giving the impression of a megacolon or Hirschsprung's Disease. However, the absence of an increased anal sphincter tone ruled out the latter.

An exploratory laparotomy revealed massive adhessions, and multiple internal intestinal fistulas, as well as a long jejunoileal atresia measuring about 15 cm in length. The fistulas were gastrocolic, jejunocolic and several other enteroenteral ones.

To avoid a time consuming dissection in isolating the individual fistulas, which could worsen the already poor condition of the infant, an en-block resection of the atresia together with a number of the fistulas was performed. This lead to preserving only about 20 centimeters of ileum. The ascending colon and about half of the transverse and the whole of the left colon. To establish a continuity of the GI-tract a Rouxen-Y gastroenterostomy and an end-to-end anastomosis of the transverse colon after closure of the gastrocolic fistula was performed.

The postoperative period was complicated by severe dehydration and further weight loss. The child died on the tenth postoperative day, possibly from water and electrolyte imbalance following malabsorption.

DISCUSSION

While autosomal recessive pattern of inheritance [1] does explain a familiar occurreace, mesenteric

Dr. David nii-Amon-Kotei

vascular accidents [7, 8, 9] and failure of recanalisation of the intestine after a period of epithelial solidification [6], intussusception (Chiari, H.(1888) Über eine intrauterine entstandene und von Darmatresie gefolgten Intussusception des Ileus ', Prag. Med. Wschr., 13, 399) and adhesions, strangulations and vascular damage following meconium peritonitis (Forshall, I., Hall, E.G., and Rickham, P.P. (1907) Meconium peritonitis, Br. J. Surg. , 40, 31) and twisting off of a segment of the intestine [9, 14,] indicate to mechanisms leading to intestinal atresia. The same, however, fail to provide a mathematically reproducible variable as relates to the structure and function of the intestine. The case presented here being associated with internal fistulae does not only add to the diversity of opinion on the etiology of intestinal atresias but also suggests primary and basic physical changes in the structure or function as the very first etiological factor which might be followed by the atresia per se as a secondary pathological change. The structural change in our case is seen as the internal fistulae formation which lead to multiplication of the channeling. Parameters of the flow of the contents of the GI-tract such as the rate and direction of flow constitute the functional changes. This on the other hand opens up the debate whether the atresia preceded the formation of the fistulas, or vice versa. The latter presupposes an intraluminal pressure build up before an obstruction and leading to rupturing of the intestinal wall when its point of elasticity is exceeded. The atresia here therefore can only be secondary, occurring in the intestinal segment after the obstruction on account of collapse due to lack of irrigation by the flow of intestinal contents. In the former, introduction of junctional points into the GI-tract leads to multiplication of direction and target sites of the flow. The subsequent and new flow directions can follow definite pathways in the form of fistulas. They may also not have a defined pathway, such as found, with perforations. Both fistulas and perforations convert the in series connection of the different parts of the normal GI-tract into a reticulation system on which principles of fluid mechanics may be applied [12]. The multiplication of the channeling brings with it corresponding multiplication of discharge or target points and the distribution of the volume rates of the contents and the total head loss or pressure difference.

In the case presented here, the clinical picture of a megacolon or Hirschsprungs Disease is seen as an increase in the supply of intestinal contents to the large intestine; this being the result of short circuiting by the internal fistulas as can be inferred from the algebraic summation of volume flow rates and head losses in a reticulation system [9,4,5,6]. The same picture can however be produced by stagnation or increase in intraluminal pressure before a resistance such as a stenosis or constriction caused by the aganglionosis in Hirschsprung's Disease. Such an intraluminal pressure increase could have also ruptured the intestine leading to perforations and formation of the fistulas.

The inability of the infant to thrive is the result of malabsorption, which was possibly brought about by the diversion of nutrients away from secretory, digestive and absorptive surfaces of the intestine. In the present case, this has had similar clinical outcome like a short bowel syndrome.

The long jejunoileal atresia can be the result of failure of irrigation, effected by the parallel diversion of the flow of the contents of the small intestine. However it is uncertain whether atresia occurred before the fistula formation. Although the present author is unable to answer the question, this case demonstrates that parallel diversions of the GI-tract Pneumoperidium secondary to benign gastric ulcer perforation [15], acute cardiac tamponade resulting from gastropericardial fistula [13] and postoperative peritonitis due to gastric and duodenal fistulas [8]; and even acute pancreatitis can also be seen as some of the complications of a parallel diversions of the GI-tract. Roux-en-Y gastroenterostomies, colostomies and ileostomies are however curative and palliative uses of parallel GIT-diversions.

REFERENCES

1. **Blythe, H., Dickson, J.A.S.:** Apple peel syndrome (congenital intestinal atresia): a family study of seven index patients. J.Med Genet, Vol. 6,Pp 275- 277, 1969
2. **Brown, J.J.M.:** Small intestinal obstruction in the newborn. Ann. R. Coll. Surg. Engl, Vol. 20, pp 280, 1957
3. **Burgler, J.:** Fluid Mechanics for Technologists. Longman, Cheshire, 1989
4. **Chiari, H.:** Über eine intrauterine entstandene und von Darmatresie gefolgten Intussusception des Ileus ', Prag. Med. Wschr., 13, 399 1888
5. **Daugherty, R.L., Franzini, J.B.:** Fluid Mechanics with engineering applications, 6th edition, McGrwahill Book Company, New York, 1965
6. **Forshall, I., Hall, E.G., and Rickham, P.P.:** Meconium peritonitis, Br. J. Surg. , 40, 31, 1907

7. **Gross, R.E.:** The surgery of infancy and childhood. W.B. Saunders, Philadelphia, 1953
8. **Gustavsson, G., Ilstrup, D.H., Morrison, P.:** The Roux-stais syndrome after gastrectomy. Am J Surg., Vol. 155, pp 450- 494, 1988
9. **Levy, E, Cugnec, P.H., Frileux, P., Hannoun, L., Parc. R., Huguet, C., and Loygue, J.:** Postoperative peritonitis due to gastric and duodenal fistulas- operative management by intraluminal aspiration: report of 23 cases. Br. J. Surg. Vol. 71, pp 543- 546, 1984
10. **Louw, J. H.:** Congenital atresia and severe stenosis in the newborn. S. Afr. J. Clin Sci, vol. 3, pp209, 1984
11. **Louw, J.H. Barnard, C.N.: Congenital atresia:** Observation of its origin. Lancet 1995:ii 1065-1067
12. **Neher, M, Kümerle, F.:** Gastrointestinale Komplikationen bei akuter Pankreatitis Dtsch. Med. Wschr. Vol.103, pp1400-1404, 1978
13. **Nicolau, N., Katz, G., Coblan, A.A.:** Gastropericardial fistula presenting as acute cardiac tamponade. Sa. Mediese Tydskrif Deel. Vol. 65, pp51- 55, 1984
14. **Nii-Amon-Kotei, D.:** Spontaneous congenital parallel shunting of the gastrointestinal tract: flow dynamic complications. Pediatr. Surg. Int. Vol. 9, pp498- 500, 1994
15. **Nixon, H.H:** Intestinal obstruction in the newborn. Arch. Dis. Childh., Vol. 30, pp13, 1955
16. **Rombilt, D.W., Alexander, J.W.:** Pneumopericardium secondary to perforation of benign gastric ulcer. JAMA Vol. 191, pp140 142, 1965
17. **Spriggs, N.I.:** Some cases of congenital intestinal obstruction, Lancet vol. I, pp94, 1910