Nigerian Journal of Paediatrics 1981; 8 (2): 40-44

Congenital Anorectal Anomalies in Western Nigeria

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Summary

Adekunle OO and Johnson AOK. Congenital Anorectal Anomalies in Western Nigeria. *Nigerian Journal of Paediatrics*, **1981; 8:40.** Forty-five patients with congenital anorectal anomalies are presented. The sex ratio is 1:1. The high (supralevator) type anomalies are slightly more common than the low (translevator) anomalies. Associated structural anomalies occurred in only 13% of all the cases. Results of treatment were good in low anomalies, satisfactory in intermediate, but very poor in high anomalies. Late presentation, high default rate after colostomy, and paucity of specialised paediatric units, appear to contribute to poor prognosis. It is suggested that avoidance of colostomy in low anomalies, and definitive treatment during the first admission in intermediate and high anomalies will improve prognosis.

Introduction

After congenital inguinal hernias, congenital anorectal anomalies appear to be the commonest congenital gastrointestinal anomaly in Nigerian children. Anorectal anomalies pose peculiar problems of management in our environment partly because of the socio-cultural beliefs and practices of the population, and partly because of the paucity of specialised centres where corrective surgery for these anomalies can be offered. A previous review of these anomalies at the University College Hospital (UCH), Ibadan, concerned twenty patients.[1] The present communication presents the study of a larger number of patients and highlights the problems of management of the condition. It also suggests a treatment policy which we believe is best suited to the realities of an unsophisticated society.

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Materials and Methods

The case records of all infants with congenital anorectal anomalies admitted into the paediatric wards of the UCH, Ibadan, between 1970 and 1977, were studied. Information collected included age at presentation, sex, nature of the anomalies, associated congenital anomalies, treatment and outcome.

Results

Sex, Age and Incidence

Forty-five patients with congenital anorectal anomalies were seen during the period covered by the study. They consisted of 22 males and 23 females giving a sex ratio of approximately, 1:1. In the same period as the study, there were 9,577 admissions into the paediatric wards and 389 of these were cases of congenital anomalies of the gastrointestinal tract. Thus the 45 cases of congenital anorectal anomalies represent 0.47% of all paediatric admissions and 11.8% of congenital anomalies of the gastrointestinal tract.

Table I shows the age at presentation. Eight patients (18%) were admitted on the day of birth and 26 (60%) presented in hospital by the

TABLE I

Age at Presentation of 45 Children with Congenital Anorectal Anomalies

Number of Patients	26	3	4	2	–	1	–	1	–	–	–	3	1	–	–	1	3
Age at presentation (Weeks)	1	2	3	4	5	6	7	8	9	10	11	12	13	14	15	16	>16

end of the first week. Eight (18%) were aged three months or above before presenting in hospital.

Types of Anomalies

These are summarised in Table II and have been classified according to the International Classification proposed by Stephens and Smith.[2]

The 19 patients with the translevator type of anomaly consisted of 8 males and 11 females. Among the males, there were 2 cases of anal stenosis, 5 of completely covered anus and 1 of anocutaneous fistula. In contrast, there were among the females, 3 cases of anal stenosis, 4 of anocutaneous fistula and 4 of anovestibula fistula. The 7 children with the intermediate type anomaly consisted of 4 females and 3 males, while the supralevator type of anomaly occurred in 11 males and 7 females. The intermediate and supralevator anomalies were associated with recto-urethral and rectovesical fistulae in males and recto-vaginal fistulae in females.

TABLE II

Types of Defects in 45 Children with Congenital Anorectal Anomalies

Type of Anomalies	*No. of Patients*	*% of Total*
Translevator (Low)	19	42
Supralevator (High)	18	40
Intermediate	7	16
*Not Known	1	2
Total	45	100

*Type of defect could not be determined because of inadequate description in the case records.

Associated Anomalies

Six patients (13%) had associated non-anorectal anomalies, and the nature of these anomalies are listed in Table III. It is noteworthy that the two associated anomalies requiring urgent surgical intervention occurred in patients with supralevator anomalies. In addition, three children with supralevator anomaly had associated neonatal jaundice; one of these had erythrocyte glucose-6-phosphate dehydrogenase (G-6-PD) deficiency and Christmas disease, while another had G-6-PD deficiency only.

Treatment and Outcome

The treatment employed in the present series of anorectal anomalies are shown in Table IV. Among the 19 patients with translevator defects, the procedures carried out consisted of anal dilatation in 2 patients, perforation of anal membrane in 3, and anoplasty in 5. The remaining 9 patients had initial colostomy and this was

TABLE III

Associated Abnormalities in 45 Patients with Anorectal Anomalies

Type of Anorectal Anomaly	*Type of Associated Anomaly*	*No. of Patients*
Supralevator (High)	Intestinal malrotation	1
	Talipes with Omphalocele	1
Translevator (Low)	Hypospadias	1
	Polydactyly	1
	Hemivertebrae, Rib defects and Undescended Testis	1
Intermediate	Atrophy of radial border of left hand	1
Total		6

followed by anoplasty in 3, abdomino-anal pull-through in 2 and perforation of anal membrane in one. All these 16 patients also had subsequent regular anal dilatation post-operatively and achieved faecal continence. Of the remaining 3 patients, one died soon after colostomy and 2 were lost to follow-up before corrective surgery could be performed. Thus 84% of children with translevator anomalies had good results after correction of the anomalies.

All the 7 patients with intermediate type anomaly had initial colostomy. This was followed by abdominoanal pull-through in 4 patients, and anoplasty in one. The remaining 2 patients defaulted after colostomy. One of the 4 patients who underwent abdominoanal pull-through died, while one developed faecal incontinence. The remaining 2 and the child who underwent anoplasty had good results giving a success rate of 43%.

TABLE IV

Treatment in 45 Patients with Congenital Anorectal Anomalies

Treatment	*Type† of Anomaly*			
	T	*I*	*S*	*U*
Dilatation only	2	–	–	–
Perforation of Anal Membrane and Dilatation	3	–	–	–
Anoplasty	5	–	–	–
Colostomy only	3	2	13	1
Colostomy and Anoplasty	3	1	–	–
Colostomy and Abdomino-Anal Pull Through	2	4	3	–
Colostomy and Perforation of Anal Membrane	1	–	–	–
No Treatment	–	–	2	–
Total	19	7	18	1

† *Types:*
T — Translevator
I — Intermediate
S — Supralevator
U — Unknown

Of the 18 patients with supralevator type of anomaly, 2 died before any form of surgical treatment could be offered, while 16 had colostomies initially. Following the colostomies, 2 more patients died while 11 defaulted from the clinic, and only 3 returned to have abdominoanal pull-through operations. Of these 3, one had good anal sphincteric action but the remaining two developed incontinence of faeces. Thus, the success rate for treatment of the supralevator anomalies was only 6%.

Discussion

The reported incidence of congenital anorectal anomalies is one in every 5,000 births and rarely is there any family history.[3] Because of the diverse nature of these anomalies, there has been some confusion in the classification, but the one proposed by Stephens and Smith[2] is now widely used. This classification, which recognises three major subdivisions of anomalies, is based on anatomical rather than embryological considerations. It consists of supralevator (high), translevator (low) and intermediate anomalies. The translevator anomalies are usually easy to treat and have good prognosis because the sphincteric mechanism is intact. On the other hand, the intermediate and supralevator anomalies are difficult to treat and almost always require initial colostomies, a social stigma in the Nigerian society. In addition, because the sphincteric mechanism is commonly deficient in these types of anorectal anomalies, the outcome after definitive surgery is often unsatisfactory.

The diagnosis of an anorectal anomaly is usually easy because of the absence of an external anal opening or the presence of an ectopic one, and hence most patients present at birth or soon after. Some patients with wide ectopic external openings which were initially adequate for bowel evacuation may present late because the opening may subsequently become inadequate as the stools become firmer. Constipation with recurrent straining prompts the mothers to seek medical attention in such cases.

The determination of the variety of anomaly is not always straightforward. Examination of the perineum will reveal most cases of translevator anomalies either as a bulge or as an ectopic anus in the perineum or as a pinhole anal opening. In the supralevator variety, the perineum is blind but meconium may be seen coming from an opening high on the vaginal wall in females, while gas bubbles or greenish coloured urine may be passed in the males. The classical Wangesteen and Rice[4] method of plain radiography of the child in the inverted position measures the distance between the position of the anal dimple and the termination of rectal/anal air translucency. However, the presence of some tenacious meconium plug at the distal end of the rectum undisplaced by air may increase this distance and give a false impression of the level and vareity of the anorectal anomaly. Lateral invertogram is therefore often more helpful.[2] In this view, the pubococcygeal line is just above the level of the puborectalis sling which is where supralevator anomalies end. More precise assessment is also possible by means of a fistulogram through the ectopic external opening or barium examination through the distal colostomy loop.

Supralevator and translevator anomalies were equally common in the present series which is in keeping with the observation by Bankole.[1] In contrast, McPherson[5] reported that supralevator varieties were more common while Partridge and Gough[6] found that the translevator anomalies occurred more commonly.

The International Classification of Stephens and Smith[2] recognises two major types of supralevator anomalies. These are anorectal agenesis and rectal atresia. The former is the commoner and usually forms a fistula with the urogenital tract in both sexes. Rectal atresia is more difficult to diagnose because the anus appears normal and there is no associated fistula. In the present series there were 16 cases of anorectal agenesis of which 9 were females and 7 were males. All the females had high recto-vaginal fistulae while 5 males had rectovesical and 2 recto-urethral fistulae. Rectal atresia occurred in 2 patients, one male and one female.

Two groups of patients with translevator anomaly are readily identifiable: those with an abnormal opening at the normal site, for example, anal stenosis and covered anus (complete), and those with an ectopic opening. In males, the ectopic opening will be found on the perineum while in the females, the opening may be in the perineum or at the vestibule. While anal stenosis and covered anus formed the bulk of the translevator varieties in males, vestibular ectopic anus was commoner in the females. The intermediate type of anorectal anomalies is the least common in this series and most of the cases are females with low rectovaginal fistulae.

The reported incidence of associated congenital defects in anorectal anomalies varies between 25% and 80%.[7–9] Only 13% of our patients had associated structural anomalies, and this figure rises to 18% if the 2 patients with erythrocyte G-6-PD deficiency are included. However, it seems likely that the true figure would have been higher if the default rate after colostomy had been less and if all the children had been more extensively investigated.

Most infants with anorectal anomalies require some form of immediate treatment. Anal stenosis needs only regular dilatation, while covered anus requires perforation of the membrane followed by regular dilatation. Other translevator anomalies with ectopic opening will require some form of anoplasty. Most cases with intermediate and all those with supralevator anomalies usually require a temporary transverse colostomy to allow time for detailed analysis of the situation, and in some patients, for immediate relief of acute intestinal obstruction. One stage definitive procedure in supralevator and intermediate anomalies have been recommended by some authors.[5] However, the view is widely held that definitive treatment should be delayed to between 6 and 12 months as earlier treatment may damage the puborectalis sling.[3] The effect of the age at definitive surgery on the outcome of management could not be

assessed in our patients because of the high default rate after colostomy.

In the present series as well as in others, the result of treatment of translevator anomalies was excellent. Sixteen of the 19 patients were continent of faeces after operation. There was one death, 5 days after colostomy. In the intermediate variety, the results were satisfactory if the two cases who defaulted after initial colostomy are excluded, leaving 3 of 5 patients completely cured. The results of the supralevator type of anomaly were the poorest. Anal continence was satisfactory in only one of the 3 patients who had pull-through operations and there were 4 hospital deaths. The poor results of treatment in supralevator anomalies in this series is in keeping with the experience from other centres.[5 6 10]

There was a high default rate after colostomy. There were 33 colostomies of which 3 died immediately post colostomy, 15 defaulted (default rate, 45%) while the rest had definitive treatment. Default after colostomy is perhaps not unexpected because of the social stigmata generally attached to this procedure in our environment. For example, a recent review of carcinoma of the rectum in Ibadan revealed that 30% of patients refused abdomino-perineal resection because colostomy was socially unacceptable (unpublished data). Majority of cases who failed to return after initial colostomy may have died either from natural causes, from associated anomalies, from intercurrent diseases or from neglect by embarrassed and superstitious parents. In this context therefore, the role of the Medical Social Worker and the Health Visitor in giving the parents of children with colostomies much needed support and guidance is of paramount importance. It may also be worthwhile to consider one-stage definitive operations in which the colostomy if any, would be closed before the child is discharged from the hospital. Major constraints against this include late presentation in many instances resulting in an acutely ill child, limited anaesthetic facilities and specialised paediatric nursing care. All these make a major operation hazardous. Besides, the great demand for the few paediatric beds available for other acutely ill children makes a long hospital stay sometimes impracticable.

Despite these reservations and in view of the immense social stigma attached to a colostomy, it is suggested that colostomy should be avoided in cases of low anomalies and definitive treatment carried out at the time of first presentation. Definitive surgery on children with high anomalies and without severe associated anomalies should not be delayed beyond the first admission. It is hoped that these measures coupled with the support and encouragment of Medical Social Workers and Health Visitors will considerably reduce the default rate and improve morbidity and mortality associated with the treatment of congenital anorectal anomalies in a developing country like Nigeria.

Acknowledgements

We acknowledge the part played by our Medical and Nursing colleagues in both the Departments of Paediatrics and Surgery in the management of these patients.

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Accepted 27 May 1980