Results of treatment in children with anorectal malformations in Calabar, Nigeria

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Summary

Leading symptoms of anorectal malformation in the neonatal period are abdominal distention, non-passage of meconium and constipation. When present, vomiting is a late symptom. In a study in Calabar, Nigeria, patients were observed to present late, and teenage mothers in rural communities were mostly affected. Female neonates were affected more than males in a ratio of 1.5:1. Classification into low and high abnormality was adopted and proved practical in terms of identification of the pathology and treatment of the lesion. Patients with low abnormality (*N* = 24, 44.4%) were treated with perineal cut-down, while those with high abnormality (55.6%) had initial palliative colostomy before a definitive abdominal perineal pull-through procedure. Faecal incontinence (13%), anal stenosis (11.1%), constipation (7.4%) and colostomy prolapse (5.6%) were noted to be associated complications.

Poverty and ignorance were noted to be the main factors affecting treatment outcome. A concerted public enlightenment campaign is therefore required.

Worldwide, neonatal intestinal obstruction is commonly caused by anorectal malformation, and in developed countries it occurs more frequently in boys than girls.[1] Reports from some centres in the tropics found anorectal malformation to be the second most common cause of neonatal intestinal obstruction.[2] In a previous report from this centre anorectal malformation was found to be the third most common cause of intestinal obstruction in children, accounting for 10.8% of cases.[3] Diagnostic difficulties as a result of its frequent association with other anomalies has made treatment of this condition difficult.[4]

Poor understanding of this condition in our environment borders on ignorance. Mandatory visits to traditional healers result in late presentation to hospital, severe dehydration and poor clinical condition.[5] To the best of the authors' knowledge, no comprehensive report on anorectal malformations in children in the region had been documented. This study was therefore undertaken to highlight the magnitude of the problem, clinical presentation and treatment outcome in our environment.

Patients and method

This was a prospective study of all patients who presented to the University of Calabar Teaching Hospital (UCTH) between January 1992 and December 2001. The UCTH is the only tertiary referral health institution serving the whole of the south-eastern axis of the country, with a population of about 7 million (1991 census). To qualify to be included in this study patients had to fulfil the following criteria: (*i*) confirmed diagnosis of anorectal malformation; and (*ii*) insertion of a clinical thermometer into the anatomical anal canal was not possible, thus differentiating the condition from other forms of neonatal obstruction.

Relevant information pertaining to the antenatal record was obtained from the parents where possible. Sex, age at presentation, the state of nutrition and hydration, remedies used before presentation at UCTH and outcome of such remedies were all noted. Questions were also asked about the parents' age and their level of education, occupation and place of residence. Treatment offered at the UCTH, outcome of such treatment and complications (if any), were also noted. Relevant clinical and laboratory examinations included general clinical assessment of the patient, radiological examination, ultrasound of the chest and abdomen (where possible), haemogram and urinalysis. In all cases treatment started with fluid replacement therapy and antibiotics. Palliative colostomy (where necessary) and definitive surgical correction were effected only after stabilisation of the clinical condition of the patient. For ease of clinical assessment and treatment of the anomaly classification of the pathology into low and high abnormality was adopted and treatment of the lesion was based on this.

Results

During the period of this study 54 children were diagnosed and treated for anorectal malformation. There were 32 girls and 22 boys, giving a female/male ratio of 1.5:1. The majority of patients presented after the first 24 hours of life, with some patients seen after as long as 72 hours (Table I).

Results of the clinical evaluation showed that low abnormality was seen in 24 patients: covered anus (8 patients), ectopic anus (6 patients), stenosed anus (7 patients), and membranous stenosis (3 patients). High abnormality was seen in 30 patients, with anorectal agenesis occurring in 18 patients, rectal atresia in 10 patients, and cloaca in only 2 patients.

Most of the parents of these patients (95%) were of low socio-economic status (artisans, peasants or labourers), with 78% of them living in rural areas. The majority of the mothers (74%) were teenagers.

Dominant clinical features were abdominal distension, non-passage of meconium, and constipation. Vomiting was rarely seen and when present it was a late sign (Table II).

Clinical evaluation of the patients revealed that in 3 cases there was concomitant cardiac anomaly, while in 2 patients the anomaly was associated with deformities of the musculoskeletal system (syndactyly of the toes). However, 2 patients had associated gastrointestinal tract (GIT) anomaly, presenting as ileal atresia. Where patients with low abnormality had termination of the bowel below the pelvic floor, perineal cutdown procedures were instituted. In the high abnormality group (30 patients) with termination above the pelvic floor, palliative colostomies were instituted for deflation, and the definitive abdominal perineal pull-through operation was deferred until adequate weight gain and general stabilisation of the patient's clinical condition was achieved.

Eleven patients with palliative colostomies were lost to follow-up and later confirmed dead, apparently due to neglect by their families.

TABLE II. CLINICAL FEATURES IN 54 PATIENTS WITH ANORECTAL MALFORMATIONS AT UCTH, CALABAR

Clinical features*	No. of patients	%
Abdominal distension	**54**	**100**
Non-passage of meconium	**41**	**76**
Constipation	**38**	**70**
Fever	**16**	**30**
Vomiting	**11**	**20**

*Some patients had more than one clinical feature.

TABLE I. AGE AND SEX DISTRIBUTION IN 54 CHILDREN WITH ANORECTAL MALFORMATION AT UCTH, CALABAR

Sex	Age (days) < 1	1 - 2	3 - 4	5 - 6	> 6	Total no. of children
Male	**5**	**10**	**6**	**1**	**–**	**22**
Female	**6**	**11**	**6**	**7**	**2**	**32**
Total	**11**	**21**	**12**	**8**	**2**	**54**

During a follow-up period of between 6 and 24 months, the following complications were noted: colostomy prolapse (3 patients), constipation (4 patients), anal stenosis (6 patients) and faecal incontinence (7 patients).

Discussion

It is difficult to determine the true incidence of any gastrointestinal anomaly in our environment since records of births in the community are not available. It is well known that the majority of Nigerian mothers deliver outside of orthodox medical facilities. Because of this we were unable to determine the incidence of congenital anorectal lesions in the present survey. The study is also unlikely to provide any accurate figure on what obtains in the community since, presumably as a result of the worsening economic situation in Nigeria, a number of affected infants might not have been brought for medical attention. Taking all these factors into consideration, our hospital figure is very likely to be an underestimation of the real situation. Nevertheless, since UCTH is the only referral centre for paediatric surgery in south-eastern Nigeria, it can be assumed that the hospital had pooled the bulk of Nigerian infants born with major GIT anomalies in the area. It is therefore hoped that the findings of this survey will provide a clue as to the pattern of congenital anorectal lesions in our environment.

The study revealed a female preponderance among the infants with congenital anorectal malformation, which contrasts with the experience in Europe where males predominate.[1] Perhaps this may be explained by the observation that in the last head count (national census 1991) females in the region outnumbered males in the paediatric age group (< 18 years).

In our series the major types of anorectal anomaly afflicting infants were classified as high abnormality (55.6%) and low abnormality (44.4%). The high type was most prevalent, which is in consonance with global experience.[1] Congenital defects have been associated with various harmful agents to which the mothers were exposed during the critical period of embryogenesis, thalidomide being a good example. In a previous study in our centre, Ekwere[6] linked penile agenesis and congenital sacrococcygeal teratoma in the population with the frequent use of insecticides. The affected population in the present survey was largely of low socio-economic status and were most likely to burn and inhale mosquito repellant coils at night or to fumigate their farms with insecticides. These agents may affect pregnant mothers, resulting in the possible birth of malformed babies. Also, it has been documented that teenage pregnancy is a major problem in our society and in attempts by these young mothers to terminate their pregnancies they ingest various concoctions.[7] Presumably, the constituents of some of these medicaments have teratogenic effects. This hypothesis constitutes a subject for further research.

A majority of patients in this study presented late for orthodox medical attention. Undue delay in seeking hospital treatment is a well-known phenomenon in our environment; a good number of the populace prefer to shop around for traditional remedies and only consult orthodox medical facilities as a last resort.[7] Ignorance, living far from a medical institution, and inability to afford hospital fees may explain this. Intestinal obstruction with delay in the passage of meconium beyond 24 - 48 hours, coupled with intolerance of feeds and vomiting can obviously render the infant very ill, dehydrated and moribund. Such infants require vigorous resuscitation before even a minor surgical procedure can be attempted. Traditional midwives and birth attendants need to be trained and well oriented so that they can identify infants at risk and advise prompt hospital consultation. It has been suggested that midwives be allowed to insert rectal thermometers in newborns so as to facilitate the early diagnosis and referral of infants with anorectal defects (course organised for traditional birth attendants by the State Ministry of Health in Calabar in 1998 — unpublished data).

Congenital GIT lesions can occur at multiple sites in one individual.[8] For instance in the present survey 2 infants were identified as having additional malformations at other sites within the gut. This underscores the importance of searching for other anomalies along the tract when one defect has been identified. Also the incidence of associated multisystemic anomalies in patients with anorectal dysgenesis can be as high as 50%, with vertebral and genitourinary malformation being the most common.[9] The present study revealed multisystemic lesions in 9% of the infants, reflecting the polymorphous nature of congenital malformation. These findings might have been utilised to formulate a syndromal diagnosis but the preliminary nature of this study precluded such a venture.

Classification[10] into low and high abnormalities appears to have simplified the decision on a surgical procedure for each patient. The practice of surgical operation under protective colostomy is useful as it can reduce the rate of postoperative complications and perhaps allow for satisfactory sphincteric function/continence and therefore excellent aesthetics.[11] Palliative colostomy allowed for adequate weight gain and stabilisation of the patients for later successful definitive abdominal perineal pull-through. Nevertheless, some families detested the initial colostomy procedure. Wearing a colostomy bag is regarded as a stigma and a bad omen in our society. This superstitious belief and negative attitude may have been responsible for the neglect, abandonment and subsequent death of 11 of the colostomy patients in this series. Nigerian society therefore needs effective health education in this regard.

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