Some Observations on Anorectal Malformations in Zimbabwe

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SUMMARY

Forty-six cases of anorectal malformations (ARM) were seen over a period of 28 months in the elective paediatric surgical service of the two teaching hospitals in Harare, Zimbabwe, of whom 20 (43,5%) were males and 26 (56,5%) were females. Twenty-two (47,8%) of the cases had a 'high' anomaly while 21 (45m7/5) had a 'low' anomaly, and the remaining 3 cases (6,5%) had a 'cloaca' malformation. Twelve (26%) of the cases were shown to have major associated congenital anomalies affecting mainly the uro-genital, musculo-skeletal or cardiovascular systems. About three-quarters of the cases were referrals from other hospitals. The apparently peculiar geographical distribution of ARM in Zimbabwe and some of the problems encountered in the management of these cases are presented and discussed.

A plea is made for the rational and skilful management of this major and challenging paediatric surgical problem in Zimbabwe.

INTRODUCTION

In a recent review of paediatric surgical problems in Zimbabwe[1] it was shown that anorectal malformations (ARM) were among the leading and challenging problems, along with

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Hirschesprung's disease and other forms of megacolon in childhood. It has also been shown[2] that ARM is a common and important congenital anomaly presenting as a surgical emergency in the newborn. While the special features of megacolon in Zimbabwe have been the subject of another recent communication,[3] this study, based on a personal series of 46 consecutive cases, seeks to present some information on the pattern of ARM in Zimbabwe, its geographical distribution and some of the problems encountered in its management, as such information has not yet been published.

MATERIALS AND METHODS

All cases of ARM referred to the elective paediatric surgical service under the care of the author at Parirenyatwa and Harare Teaching Hospitals, during the 28-month period, February 1984 – June 1986, have been included in the study.

The case records of all the affected patients have been reviewed and analysed to determine the following information:

(i) The age and sex distribution
(ii) The pattern of anorectal malformations
(iii) The presence and pattern of associated congenital anomalies
(iv) The place of birth and/or referral
(v) The outcome of treatment or evaluation
(vi) Special problems encountered in the management of the cases.

All the ARM cases in (ii) were classified into 'high', 'low' or 'cloaca' categories on the basis of clinical findings (including 'examination under anaesthesia' when indicated) or on the basis of the case histories submitted by the referring hospital in those patients who had surgical treatment elsewhere, in accordance with the International Classification of ARM of 1970.[4] No radiographic methods or other more sophisticated investigations were used. 'Intermediate' anomalies were categorized as 'high' and managed as such.

The outcome of treatment (v) was designated as good, fair or poor, relating to 'normal faecal control', 'occasional faecal soiling' or 'frequent faecal soiling or total incontinence', respectively, as stipulated by Nixon *et al.*,[5] in the 24 patients who have completed treatment under the care of the author and have been followed up for a period of three months to two years.

RESULTS

Forty-six consecutive cases have been referred to the elective paediatric surgical service in Harare, under the care of the author, over the 28-month period ending in June 1986.

All but 6 of the cases were referred as newborn or infants, and 11 were seen by the author before any surgical treatment had been undertaken. The late cases were aged three–nine years and had preliminary or definitive surgical treatment performed elsewhere before referral. Twenty (43,5%) of the 46 cases were males and 22 (47,8%) of them had a 'high' anorectal malformation. Details of the pattern and sex distribution of the ARM cases are shown in Table I.

TABLE I – *The pattern and sex distribution of 46 ARM cases in Zimbabwe*

Type	Male	Female	Total	%
High	19	3	22	47,8
Low	1	20	21	45,7
Cloaca	0	3	3	6,5
Total	20	26	46	100,0
%	43,5	56,5		

Twelve (26%) of the 46 cases had associated major congenital anomalies affecting mainly the urogenital, musculo-skeletal, or cardiovascular systems (Table II) which were detected either on clinical or radiological examination. Skeletal X-rays were done on those patients on whom there was a clinical suspicion of a bony abnormality and intravenous pyelography (IVP) was done only on patients with a 'high' anomaly as part of their pre-operative assessment. Associated urogenital anomalies included (1) bilateral renal agenesis; (2) urethral stricture with bilateral hydronephrosis and hydroureters, and undescended testicles; (3) unilateral renal agenesis with hypospadias; (4)

double vagina ; and (5) urogenital sinus. Skeletal abnormalities were mainly those of the sacral vertebrae (sacral deficiency), the thoracic cage and spine (scoliosis), and one unilateral congenital talipes equinovarus. One of the cases with cardiovascular anomalies had Fallot's tetralogy and the other had a ventricular septal defect.

TABLE II – *Associated congenital anomalies in 12 ARM cases*

Affected system	Number of cases
Urogenital	9
Musculo-skeletal	8
Cardiovascular	2
Alimentary	2
Central nervous	1

Table III shows the geographical distribution of 31 of the patients for whom the locality of birth or referral could be ascertained, and demonstrates that about two-thirds of these patients were from the Masvingo, Midlands and the two Matabeleland provinces, which account for less than a third of Zimbabwe's population, while the remainder came from the other four provinces.

TABLE III – *Geographical distribution of 31 ARM cases*

Area/province	All types	High types
Bulawayo (Matabeleland)*	12	8
Masvingo*	6	1
Midlands*	3	2
Manicaland	1	1
Harare	6	2
Mashonaland	3	3
Total	31	14

*Within the same contiguous area of southern and western Zimbabwe

The early results of treatment in the 24 cases who have been followed up for three months to two years since completion of treatment are shown in Table IV. All cases with a 'high' anomaly had a preliminary colostomy in the neonatal period followed by an abdomino-perineal pull through anorectoplasty at one–two years of age (preferably weighing 10 kg or more), while the cases with a 'low' anomaly had either a 'cut-back' anoplasty or a 'transfer' anoplasty, with or without a preliminary colostomy. There were no post-operative deaths.

TABLE IV – *Early results of treatment of 24 ARM cases*

Type of anomaly	Total cases	'Good'	'Fair'	'Poor'
High	8	6	1	1
Low	16	14	2	0

Of the 46 cases 2 died before surgical treatment could be undertaken; a male neonate from acute renal failure (due to presumed bilateral renal agenesis), and a four-month-old female infant from cardiac arrest during general anaesthesia prior to a surgical procedure. Four cases have been lost to follow-up before completion of treatment, and four others had had unsuccessful surgical treatment elsewhere and could not be helped by further surgery. There are 12 cases still awaiting further treatment.

Some of the problems encountered in the management of the ARM cases are illustrated in Table V. All the five examples had preliminary or definitive surgery performed elsewhere before they were referred, and have had complications arising from either failed or incorrect surgical treatment or poor follow-up care. Two of the cases could not be helped by further surgery, while the remainder still await further evaluation for possible but difficult corrective surgery.

Table VI shows the comparative pattern of ARM cases in Zimbabwe and Tanzania, taken from similar personal series of the author. While there is an overall male predominance in Tanzania, the reverse is true in Zimbabwe, but in both countries most of the males have 'high' anomalies while most of the females have 'low' anomalies. The fact that the 46 Zimbabwean cases were seen over a 28-month period while the 55 Tanzanian cases were seen over a 65-month period implies that ARM is commoner in Zimbabwe.

TABLE V – *Some ARM 'disaster' cases*

Patient	Referral reason	Management/outcome
1. NS (F7)	Post-surgical incontinence	i) Unsuccessful pena procedure ii) Terminal colostomy
2. CS (M9)	i) Post-operative anal stenosis with perianal fistulae and sepsis ii) Gross secondary megacolon	i) Control of sepsis ii) Terminal colostomy
3. HC (M6)	(i) Septic laparotomy for ?right hydronephrosis ii) Anal stenosis with gross secondary megacolon	i) RT colostomy + anal dilation and anoplasty ii) Awaiting further evaluation
4. TM (M2)	Imperforate anus with sigmoid colostomy	i) A-P anorectoplasty complicated by rectal stricture and persistent R-U fistula ii) RT colostomy, awaiting further evaluation
5. MN (F4)	i) ?Post-surgical anorectal stricture ii) Gross secondary megacolon	i) RT colostomy ii) Awaiting further evaluation

TABLE VI – *Comparative pattern of ARM cases in Zimbabwe and Tanzania in percentages*

	Males	Females	High	Low	Cloaca
Zimbabwe (46 cases)	43,5	56,5	47,8	45,7	6,5
Tanzania (55 cases)	54,5	45,5	36,4	61,8	1,8

DISCUSSION

Anorectal malformations (ARM) have always posed a major challenge to paediatric surgery, a fact which led the eminent American surgeon WJ Potts to state that 'Atresia of the rectum is more poorly handled than any other congenital anomaly of the newborn', having earlier declared that 'A properly functioning rectum is an unappreciated gift of the greatest price'.[6] The successful management of ARM requires among other things, good judgement, considerable skill and experience as well as diligence, in order to save the child with ARM from a lifetime of misery and social seclusion or worse consequences.

Despite the continuing controversy regarding the classification of ARM and the criteria for evaluating the results of treatment, the author, along with many other paediatric surgeons the world over, accepts and follows the 1970 International Classification of ARM[4] as the best diagnostic and prognostic guide in its management, and uses the criteria stipulated by Nixon and his colleagues[5] as the most practical means of evaluating the results of treatment, rather than the cumbersome 'Kelly's Code of Continence'.[7] A more recent modification of the International Classification of ARM, utilizing a variety of sophisticated investigations, and a new grading system for the results of treatment[8] offer a more scientific approach to the management of ARM but are not yet widely in use in clinical practice.

The current study was undertaken primarily because the author wished to provide some information on the situation of ARM in Zimbabwe hitherto not available in the literature, and to promote awareness among surgeons and

developing countries on the importance of correct preliminary as well as definitive surgical treatment of ARM.

It is the author's policy that for 'high' ARM and other categories in which staged treatment is indicated, a right transverse colostomy is the best form of preliminary surgery, especially when there is actual or impending large bowel obstruction, as has been the experience of Louw and his colleagues.[9] Thereafter, and in appropriate cases, the definitive objectives of a continent and cosmetic arorectum—as in all ARM cases—can be achieved by an abdomino-perineal anorectoplasty for 'high' anomalies at the age of one to two years (and a weight of 10 kg or more), just as it is possible to do so by other techniques, including the latest procedure of posterior sagittal anorectoplasty of De Vries and Pena.[10] That satisfactory results can be achieved even with limited medical resources is evident from the present results which are comparable to those from more developed countries.[5,7,11,12] On the other hand, poor or unsatisfactory results are inevitable when incorrect treatment is undertaken, or when the follow-up care is inadequate or inconsistent, and such results may be difficult or impossible to improve upon by further corrective surgery, for which there are very limited options.

The pattern of ARM in Zimbabwe is interesting in that it seems to differ considerably from that in Tanzania and elsewhere[1,5,13] in having a higher overall female predominance as well as a higher ratio of 'high' to 'low' anomalies. The apparently peculiar geographical distribution of ARM in Zimbabwe, by which most of the cases seem to come from Matabeleland and surrounding areas (Table III) is also interesting as well as puzzling, especially as the reverse pattern of distribution appears to be the case with regard to Hirschsprung's Disease (HD) which seems to be commoner in the Mashonaland areas. Whether this unique geographical feature of ARM *vis-a-vis* HD is a true reflection of the relative incidences of these disorders, and is due to genetic or environmental factors (or both), remains to be seen.

The pattern of associated congenital anomalies is similar to that seen elsewhere[11] and complies with the well-recognized Vater or Vacterl 'syndrome' or 'association'[14] although none of the 46 cases had associated oesophageal atresia or tracheo-oesophageal fistula. However, the low (26%) yield of associated anomalies was due to the selective policy of investigation and could have been much higher if all the 46 cases were thoroughly investigated.[13]

CONCLUSION

Anorectal malformations are a major and challenging paediatric surgical problem in Zimbabwe and this problem requires a uniform and rational policy of management in order to achieve satisfactory results. This would be considerably enhanced by the initial contralization of the definitive treatment of all ARM cases, in view of the limited availability of resources and expertise at present.

The apparently peculiar geographical distribution of ARM and its true incidence in Zimbabwe require further study.

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