EVALUATION OF SACRAL RATIO AS A PROGNOSTIC FACTOR IN PATIENTS WITH ANORECTAL MALFORMATIONS

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Abstract- Correlation between sacral ratio and bowel function as well as fecal continence has been well documented in patients with anorectal malformations (ARMs). One hundred and twenty children with ARMs were investigated in this study. Sacral ratio (SR) was measured from pelvic X-rays of patients. Among these, 52 patients (43%) had no pelvic X-ray and were excluded from this study. SR was measured by drawing three horizontal lines, through iliac crests (A), tip of coccyx (B) and inferior point of sacroiliac joints (C). The SR was determined by dividing the distance between lines B and C to the distance between lines A and B. In the normal and well developed children, the average ratio is ≥ 0.74. The SR was higher than 0.70 in 12 (17.7%) children and less than 0.69 in 56 children (82.3%). Among children with ARMs and abnormal SR, 38 cases (68%) had SR of 0.50-0.69; 12 cases (21.5%) had SR of 0.40-0.49 and 6 cases (10.5%) had SR of 0-0.39. In children with ARMs and normal SR, the fecal incontinence was observed in 2 cases (16%). In contrast, 16 cases (29%) with ARMs and abnormal SR had functional disturbance, either fecal incontinence or soiling ($P < 0.12$). When the patients had an absent sacrum, they had zero possibility for bowel control and frequently had major urinary problems. The sacral feature and SR appear to have a direct influence on the final functional outcome in ARMs. The abnormal SR < 0.7 correlates with poor bowel function. In patients with ARMs, sacral segment and SR are important factors in post operative bowel function disturbance.

Acta Medica Iranica, 43(2): 143-146: 2005

Key words: Anorectal malformations, sacral ratio, incontinence

INTRODUCTION

The sacral ratio (SR) was proposed by Alberto Pena in 1995 as a reliable tool to evaluate sacral development in anorectal malformations (ARMs). The SR is obtained by comparing sacrum size with fixed bony parameters of pelvis. In previous studies, the average normal SR has been measured to be 0.74 in anteroposterior view (APSR) and 0.77 in lateral view (LSR).

Anorectal malformations are among the most common anomalies of the gastrointestinal tract, with an incidence ranging between 1:1500 and 1:4000 or 1:5000 live birth (1,3).

ARMs are a group of congenital anomalies characterized by different degrees of complexity, depending on possible involvement of muscular and nervous structures in the pelvic region. Bony abnormalities of spine range in incidence from 30% to 44%, but patients with a high lesion are more likely to be affected (48% to 54%) than those with a low lesion (15% to 27%) (4-6). There is a good correlation between numbers of sacral vertebra (the degrees of sacral anomaly) and fecal continence (the functional prognosis of the patients with ARMs), The main importance of evaluating sacral development is that the outcome and prognosis especially for fecal continence could be estimated and discussed to the parents before any surgical intervention (8-11).

Received: 4 Sep. 2001, Revised: 6 Oct 2002, Accepted: 10 Mar 2003

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MATERIALS AND METHODS

In this study we evaluated 68 patients with ARMs whom were operated upon in the Children's Medical Center of Tehran University of Medical Sciences from 1989 to 1999. We obtained informed consent from all parents.

Required informations were obtained from charts, parents questioner or by interview. The SR was obtained by measurement of antero-posterior (AP) radiographs of the patient's pelvis. The SR was measured by the following method:

1- The most cephalad point on the left and right iliac crests was identified. A horizontal line was drawn through these points perpendicular to the vertical axis (line A).

2- The tip of the coccyx was identified. A horizontal line was drawn through the tip of coccyx perpendicular to the vertical axis (line B).

3- The inferior (most caudal) point at the left and right sacroiliac joints was identified. A horizontal line was drawn through these points perpendicular to the vertical axis (line C).

4- The A and B and C lines should be parallel to each other. SR was calculated by the following formula:

SR = vertical distance between the line through the sacroiliac joint and the line through the tip of the coccyx / vertical distance between the line through the top of iliac crests and the line through the base of the sacroiliac joint (BC/AB).

The SR in normal children is about 0.74 in an AP pelvic X-ray, and 0.77 in a lateral pelvic film. Children with ARMs may show ratio that vary from almost 0.00 to 1.00. Every child with ARMs and SR less than 0.40 had a lumbosacral MRI investigation for detecting a tethered spinal cord.

All children were assessed for fecal incontinence, soiling and constipation according to the International definition and classification.

RESULTS

Among 68 patients in this study, 38 (55%) were male and 30 (45%) were female; 33 children (48%) had low and 35 (52%) had high ARMs.

Musculoskeletal anomalies were found in 19 (28%) patients. Urogenital anomalies were observed in 23 (34%). Cardiovascular anomalies were observed in 8 (12%) of patients. The lowest range of associated anomalies were the concomitant gastro-intestinal anomalies (n=5, 8%).

The surgical reconstruction techniques varied between the children. From these, 33 cases (48%) were operated by posterior saggital anorectoplasty technique (PSA), and 14 cases (21%) were operated by perineal anoplasty technique (such as cutback transposition), 9 cases (13%) were operated by Stephan Sherli technique and finally 12 cases (18%) were operated by combined techniques.

The SR was calculated in 68 patients with ARMs. It was higher than 0.70 in 12 cases (17.7%) and less than 0.69 in 56 cases (82.3%). In the children with ARMs and abnormal SR, 38 cases (68%) had SR of 0.50-0.69 and 12 cases (21.5%) had SR of 0.40- 0.49 and 6 cases (10.5%) had SR of 0-0.39. On the other hand, voiding dysfunction was observed in 47 cases (84%) with abnormal SR and in 1 case (8%) with normal SR. All patients with SR less than 0.40 had fecal incontinence as well as voiding dysfunction. Lumbosacral magnetic resonance imaging (MRI) was performed in 6 children with very low SR (< 0.40), and confirmed tethered cord. The post surgical untethering confirmed some improvement of urinary and bowel function in 2 cases.

Disturbance of bowel function was observed in 16 cases (29%) with abnormal SR and 2 children (16%) with normal SR. The most common bowel dysfunction occurred in children operated by PSA technique (40%) versus 12% in children operated by Stephon Sherly technique. Urinary dysfunction was more common in children operated by Stephan Sherly technique.

DISCUSSION

In the review of the literature, the functional result after ARMs reconstruction is very variable (8) and there is no generally accepted method for assessing bowel function.

Assessments of long term results have been made

in different ages by different authors. A reasonable time to perform functional assessment during childhood is the age when normal children are toilet trained that is usually at about 3 years (11).

Clinical assessment is mainly based on history and precise detection of the child bowel habits, soiling, constipation and fecal incontinence. This is the most common method for evaluating the child's bowel functional outcome (3). Other methods such as monometric assessment, sphincter electromyography and imaging studies are also used for evaluating children with a history of ARMs.

The sacral evaluation (SR measurement) is a reliable method for assessment of bowel functional outcome (2). Sacral features appear to have a direct influence on the final functional outcome. Patients without a sacrum have no possibility of developing bowel and urinary control (3). Patients with a normal sacrum, on the other hand, are not guaranteed to have normal bowel control, but certainly have much better prognosis.

It has been assured that the number of sacral vertebrae correlates with the final functional outcome in these children very well. However, it is often difficult to count the number of sacral vertebrae accurately, particularly in patients with a very dysmorphic sacrum with fused vertebrae or hemi vertebrae. Thus SR measurement is a simple method for evaluation of the sacrum. The normal SR is $\geq$ 0.74, and the lower SR seems to be correlated with bowel and urinary dysfunction.

The SR measurement has recently been correlated with different diseases. The correlation among SR and nonresponder primary nocturnal enuresis, persistent primary nocturnal enuresis and vesicoureteral reflux in ARMs has been reported recently in the literature (13-16).

Age seems to be a possible variable factor for SR. The SR tends to increase with growth in normal population, while it decreases in some patients with ARMs (17).

The sacrum itself is a good predictor of outcome. Patients with a normal sacrum are much more likely to be fecally continent. Patients with a hypodeveloped sacrum are much more likely to be incontinent. SR has been proposed to obtain an objective assessment of the sacrum.

A hypodeveloped sacrum is also a good predictor of associated spinal problems, such as tethered cord. A child's outcome can be predicted more accurately with the knowledge obtained from a large series of patients. Parents can be realistically informed of their child's potential for bowel control, even in the newborn period. This avoids a great deal of frustrations later in life. Early establishment of functional prognosis is vital to avoid raising false expectations in the parents. Once the diagnosis of the specific defect is established, the functional prognosis can be predicted. Factors such as the status of the spine, sacrum, and perineal musculature affect the counseling of the parents. If the patient's defect is the type that suggests a good prognosis, such as vestibular fistula, perineal fistula, rectal atresia or imperforate anus without fistula, one should expect that child will have voluntary bowel movements by the age 3. Such children need supervision to avoid fecal impaction, constipation, and soiling. If a patient's defect indicates a poor prognosis, such as a high cloaca (common channel > 3 cm) or a recto-bladder neck fistula, the parents should be informed of the probability that child will need a bowel management program to remain clean, which should be implemented at the age 3-4. Careful regular follow up is necessary in these patients to reassess their prognosis accurately and to avoid problems that can dramatically impact their ultimate functional results.

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