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Bone health in patients with cloacal exstrophy and persistent cloaca after bladder augmentation

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Abstract

Purpose: In this study, the prevalence of osteoporosis was evaluated in patients operated on for cloacal exstrophy or persistent cloaca and who had undergone bladder augmentation.

Methods: Seven patients operated on because of cloacal exstrophy and 3 patients operated on because of persistent cloaca were evaluated. Patients were clinically assessed. Areal bone mineral density for the lumbar spine (L1-L4), femoral neck, and whole body was measured with dual-energy x-ray absorptiometry. Acid-base balance, plasma 25-hydroxyvitamin D (vitamin D), and gonadal function were determined and correlated with dual-energy x-ray absorptiometry results.

Results: The patients were small in height (median, −2.2 SD). According to areal bone mineral density measurements, 40% of the patients had significantly reduced BMD. When the values were adjusted for patient size, only 1 patient had slightly reduced bone mass. Half of the patients had low vitamin D values. The height-corrected bone mineral content values showed a positive correlation with vitamin D values ($P = .0072$) but were not different in patients with and without hypogonadism ($P = .33$).

Conclusions: Patients with cloacal exstrophy or complicated cloaca have many risk factors for osteoporosis. However, this study showed good overall bone health in affected adolescents.

Cloacal exstrophy is a rare and complex malformation that includes omphalocele and 2 exstrophic bladders, between which there is an open cecum and blindly ending microcolon. A persistent cloaca on the other hand is defined as the confluence of the rectum, vagina, and urethra into a single common channel. Spinal malformations such as lipomeningocele and tethered cord are common in all these patients. Cloacal exstrophy was uniformly a fatal condition until 1960 [1]. Nowadays, most of the patients have colostomy and augmented bladder [2]. In the case of persistent cloaca, bladder augmentation is only rarely needed. Although most of the patients with cloacal exstrophy are genetically males, most of them have been raised as females because of rudimentary phallic structures [2]. Patients with persistent cloaca are always raised as females.

Most of the reports dealing with cloacal exstrophy focus on problems commonly faced in the care of these patients: incontinence, sex determination, problems related to intestinal function, and weight development [3]. Many chronic diseases and conditions such as inflammatory bowel disease and bladder augmentation may have adverse effects on skeletal development and bone mass accrual and thus result

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in osteoporosis [4,5]. Osteoporosis is the most common bone disease in the Western world. It is characterized by reduced bone mineral density (BMD) and increased risk of fractures [6]. Early diagnostic workup and screening for osteoporosis have been recommended for patients with risk factors [6]. Patients with cloacal exstrophy and complicated cloaca may have many risk factors for osteoporosis, such as decreased physical activity, gonadal dysfunction, nutritional problems caused by short bowel syndrome, and bladder augmentation. In this study, we assessed a group of 10 patients with cloacal exstrophy or persistent cloaca for various parameters of bone health.

1. Patients and methods

1.1. Patients

Seven consecutive patients with cloacal exstrophy and 3 patients with persistent cloaca and intestinal bladder augmentation aged 9 years or older and followed by us at Helsinki University Hospital, Helsinki, Finland, were assessed in 2004 to 2006 for various characteristics of bone health as part of their regular follow-up program. Patients with cloacal exstrophy ranged in age from 9.4 to 21.0 (median, 13.9) years, and the 3 patients with persistent cloaca and bladder augmentation were 10.3, 16.8, and 25.8 years of age. The cloacal exstrophy group comprised 1 male and 6 females. Four of the female patients had had sex reassignment from male to female with bilateral orchiectomy during the neonatal period. The 3 patients with cloaca were by definition females. All 10 patients had abnormal spine (4 lipomeningocele, 3 tethered cord, 2 caudal regression, 1 vertebral malsegmentations and scoliosis). Six of 7 patients with coacal exstrophy had an end-type intestinal stoma (3 ileum, 3 colon). Also, one patient with persistent pouch colon–type cloaca had an intestinal stoma. All 10 patients had bladder augmentation at a median age of 7.5 (range, 0.6-12.6) years or 3.1 to 13.7 (median, 9.4) years before the study assessment. Augmentation material was stomach in 4, colon in 3, combined ileum and colon in 2, and ileum in 1 patient. One patient had bicarbonate medication because of constant acidosis associated with colocystoplasty. In addition, one patient with gastrocystoplasty and tendency to metabolic alkalosis and hypochloremia had NaCl and KCl substitution. Two adult patients had been prescribed estrogen replacement owing to postgonadectomy hypogonadism, but neither of them was using it. One patient with cloaca used progesterone to prevent pelvic endometrioma formation.

2. Methods

Hospital records were reviewed for clinical data. All patients were clinically assessed, and weights and heights were measured. Heights were compared with Finnish normative data and expressed as *Z* scores (deviation, in SD units, from the mean value for age and sex) [7]. Body mass index (BMI) was calculated as body weight divided by the height squared (kg/m^2). Areal BMD (aBMD) for the lumbar spine (L1-L4), femoral neck, and whole body was measured with dual-energy x-ray absorptiometry (DXA, Hologic Discovery A, Bedford, MA). The aBMDs (lumbar spine, femoral neck, and whole body) were transformed into *Z* scores by comparing the values with equipment-specific reference values. The *Z* score is the number of SDs a given BMD measurement differs from the mean for age- and sex-matched reference population. *Z* scores less than −2.0 were regarded as significantly subnormal. As most of the patients had short stature, whole-body bone mineral content (BMC) was compared with lean tissue mass (LTM) and corrected for height, as presented by Högler et al [8]. Bone mineral content and LTM were both measured by the same DXA equipment. To identify possible vertebral compression fractures, an anteroposterior and lateral image of the thoracic and lumbar spine (instant vertebral assessment) was obtained with DXA using the same scanner as for BMD measurements [9].

Blood tests included plasma calcium, plasma phosphate, plasma alkaline phosphatase, plasma parathyroid hormone, 25-hydroxyvitamin D (S-25-OHD), plasma follicle-stimulating hormone (P-FSH), plasma luteinizing hormone (P-LH), testosterone, estradiol, as well as venous standard bicarbonate and base excess. Vitamin D deficiency was considered severe if S-25-OHD was below 20 nmol/L and moderate (ie, hypovitaminosis D) if S-25-OHD was between 20 and 37.5 nmol/L [10]. The patient was considered to have acidosis if venous base excess was less than −2.5 and venous standard bicarbonate was less than 22.

3. Results

The patients' median height *Z* score at the time of the study was −2.2 (range, −4.5 to −0.4); it was below −2.0 in 7 of 10 patients. The median height *Z* score at the time of augmentation was −2.9 (range, −5.0 to −0.6); no significant changes in height *Z* score or growth velocity were observed in any of the patients when comparing the growth data before and after augmentation. The median height-adjusted weight at study assessment was +6% (range, −17% to +22%), and the median BMI was 16.9 kg/m^2 (range, 15.0-22.2 kg/m^2).

Blood tests showed in one patient, who had previously undergone gastrocystoplasty, acidosis caused by excess NaCl and KCl supplementation but normal acid-base balance in the others. Five patients (50%) had an S-25-OHD concentration consistent with vitamin D deficiency (severe deficiency in 3 and moderate deficiency in 2). Plasma calcium, -phosphate, -alkaline phosphatase, and -parathyroid hormone values were normal in all patients.

Skeletal deformities prevented the evaluation of lumbar spine, femoral head, and whole-body aBMD in 2, 1, and