

Pygomelia with genitalia duplication at Treichville Teaching Hospital: two new cases

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Pygomelia is a rare deformity (1/100 000) defined by the presence of one or more supernumerary members in the pelvic region. In this article, we report two new cases, including one that was successfully operated upon in our unit. The first case was characterized by the presence of two additional lower limbs positioned in the hypogastric region and a third vestigial epigastric upper limb associated with visceral deformations. Surgical treatment was successfully accomplished. The clinical examination of the second case objectified the third lower limb at the level of the left buttock with a rudimentary pelvis, a sacrococcygeal teratoma, and a genital duplication. The newborn child died during the investigations. The

nosological limits of pygomelia are still poorly specified because it is similar to cases of duplication of the lower limbs and dipygus. *Ann Pediatr Surg* 13:52–55 © 2017 Annals of Pediatric Surgery.

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Introduction

Pygomelia is a rare malformation (1/100 000 birth) characterized by the presence of one or more supernumerary members in the pelvic region [1]. Pygomelia poses a nosological problem because it is located on the border of four other malformations: sacrococcygeal tumors, duplications of the lower limb, double monsters, or dypigies divided into asymmetric splitting of a single embryo or two isolated adjoining embryos with one that would present an incomplete development.

The aim of this article is to report on two new cases of pygomelia that occurred in our department of pediatric surgery, to add them to the literature.

Case history

Case 1

A 1-day-old female infant born vaginally, whose 1-year-old older brother had died of unspecified causes, was born with four legs. Out of these limbs, two were quite normal and normally located, whereas two were of normal size but located in the hypogastric region (Fig. 1). We noted the presence of an upper limb in the epigastric area in addition to the two normally located (Fig. 1). Moreover, there were two vulvas side by side with a double bladder and an omphalocele type 1. The radiography of the pelvis indicated a supernumerary rudimentary pelvis. The abdominopelvic ultrasound and computed tomography also displayed a double bladder and a supernumerary colon. The cardiac ultrasound showed a patent ductus arteriosus and ventricular septal defects. Surgical excision of the limbs, pelvis, vulva, bladder, and supernumerary colon (Figs 2–5) was performed by day 43 of life through progressive dissection. The treatment for the omphalocele was achieved in the same operation (Fig. 6). At 4 years, the child is healthy and walking normally (Fig. 7).

Case 2

A newborn male infant without any particular history was presented on day 7 of life. The child was born by cesarean section. Physical examination revealed a medium general state with two normal lower limbs and a third limb on the left buttock (Fig. 8). This lower limb was immobile with a well-developed thigh, a hypoplastic leg and an equinovarus club foot. The newborn also had a sacrococcygeal teratoma infected with type I, two penises, four hemiscrotums, and two anal openings producing stools (Fig. 9). The lower limb radiography showed a rudimentary pelvis with a neoarticulation of the third limb. The abdominopelvic scanner showed a unique dysplastic kidney and dysplastic colonic duplication. Laboratory tests showed an increase in alpha-fetoprotein to 288.3 ng/ml, an initial creatinine level of 6.9 mg/l, and urea of 0.16 g/l.

Echocardiography revealed no particular abnormality. On day 11 of hospitalization, the newborn developed hypothermia and polypnea, and died after cardiorespiratory arrest.

Discussion

Some authors define pygomelia as the development of one or more supernumerary members, which can be more or less complete, and appended to the iliac crest or suprapubic region [1–4]. Pygomelia is a malformation in which nosological boundaries are poorly specified because it is closer to lower limb duplications and dypigus. Considering the rarity of this phenomenon in the human race, there are very few studies and very often the terms 'pygomelia', caudal duplication, and dypigies are confused in the literature [5]. The Anglo-Saxon term 'caudal duplication' is somewhat inappropriate in some cases of pygomelia, as one can find the presence of an upper limb at the pelvic side. This was the case of the first Ivorian observation [2]. In our observations only the second case can be considered the 'caudal duplication' of Anglo-Saxon as we found all the elements inherent to this term, such

Fig. 1



First case of pygomeia.

Fig. 2



Standard radiography of the first case.

as a supernumerary limb implanted on a rudimentary pelvis and duplication of several organs below the level of the umbilicus (external genital organs, colon, and anus).

Fig. 3



Two vulvas.

Fig. 4



Excision of the supernumerary colon.

Although our first observations revealed a set of elements that gave a closer suspicion of caudal duplication, the actual case was very different. Indeed there was an upper limb implanted in the epigastric region. This first observation is more similar to a double polymeles monster than to pygomeia. Polymeles is considered a double monster with supernumerary limbs whose implantation side is not defined and therefore could be above or below the umbilicus [6].

Pygomeia is a very rare anatomical curiosity. In fact, in 40 years of pediatric surgical practice in our country, we have encountered only three cases of pygomeia. The first case in the national literature was described exactly 17 years ago by Bankole Sanni *et al.* [2]. This was followed by our previous two cases [2]. This disease has been rarely reported in worldwide literature (1/100 000 births) [1]. In rural Africa, it is considered to be an evil spell [5]. This could cause an abandonment or even murder of this newborn in certain villages, which could explain the fact

Fig. 5



Specimen after excision.

Fig. 6



Postoperative image.

that these malformations are rarely seen by health specialists in the tropics. In Africa, the literature revealed reports of the case in Senegal, the Niger, Cote d'Ivoire, and South Africa [2,5,7,8]. Many factors have been implicated as contributing reasons for such malformations: trophic factors, traumatic, teratogenic substances,

Fig. 7



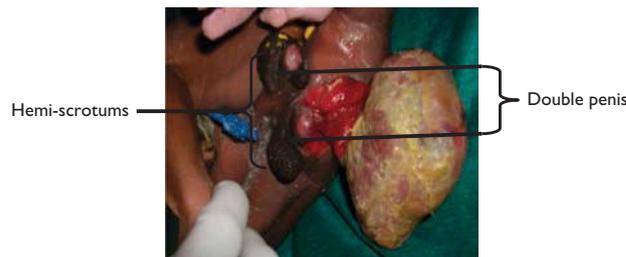
Child at 1 year of age.

Fig. 8



Second case of pygoma with a hypoplastic supernumerary limb.

Fig. 9



External view of the duplicated genital organ.

and genetic factors [9]. In our two cases none of these factors could be identified. The prognosis for these children depends on the associated malformations because pygomeia per se does not establish the prognosis [5]. In our two recent cases, one was successfully operated upon after 4 years and gave a satisfactory cosmetic and functional prognosis. The Nigerien and Senegalese cases were successfully operated upon but there was functional disability of the lower limbs in the Senegalese case probably due to the associated myelomeningocele. In these cases there was an absence of malformation, enabling one to arrive at an immediate vital prognosis. A progressive and meticulous dissection of these supernumerary limbs supervised by a good preoperative, perioperative, and postoperative care would guarantee a satisfactory result. The second case died before surgery. We highlight the fact that this latter case presented with a sacroiliac tumor that was ulcerated and superinfected (a probable sacrococcygeal teratoma) and a single kidney dysplasia. These factors may likely have contributed to an immediate poor prognosis, due to toxic shock and/or renal insufficiency, thus explaining the early death of this newborn.

To provide an addition to the literature on a national, regional, and international level, we present two new cases of this rare disease, as well as our experience of its treatment.

Acknowledgements

Conflicts of interest

There are no conflicts of interest.

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