A Case Of Dipygus Treated Surgically

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Citation

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Abstract

We report a case of caudal duplication in a 5 month old Senegalese girl. She presented a surnumerary femur articulating with a surnumerary pelvis and a rudimentary knee. This case of dipygus was treated surgically. However the child did not walk up to the age of 3 because of a sacral abnormality.

INTRODUCTION

The dipygus is a congenital distal abnormality defined as: " the presence of one or more arms between the two normal arms or buttoks of the individual ". It is an extremely rare malformation. Only 14 cases worldwide have been reported in the literature. We report a Senegalese paediatric case which was treated surgically.

CASE REPORT

N.S, is a baby girl born through normal vaginal delivery to a 15 year old primiparous mother. She was born with a waist abnormality which was hidden by her parents until the age of 5 months. Physical examination showed a surnumerary thigh of 16cm located in the sacrococcygeal region (figure 1).

Figure 1

Figure 1: surnumerary leg

The implantation circumference was 19 cm. The joint with the sacrococcygeal region was so mobile that it could extend the thigh to the back or to the popliteal region. The standard axial X- rays showed a femur articulating in a form of acetabulum which also articulated with the sacrum through a left ilium. At the distal end of the femur there was a rudimentary joint representing the knee joint followed by a proximal end of a tibia measuring 3cm. X-ray of the spine showed a myelomeningocele. The arteriograph revealed the blood supply to be from the abdominal aorta 1cm before its bifurcation. Ultrasonography also showed a myelomeningocele.

A sacro-iliac desarticulation was performed by periostal elevation of the surnumerary ilium up to the sacrum. This was done after ligation of blood vessels and one nerve trunk probably representing the crural nerve. The removed leg weighed 450g.

The pathology report noted a femoral pedicle, a complete iliac crest, a coxo-femoral joint, a head of femur, a dysplastic acetabulum, a complete femur articulating with a patella and a rudimentary tibia making a hardly mobile knee joint. There was no fibula or foot.

The post operative period was uneventful. The wound healed at day 10 post op. The child was seen for follow up at the age of three. She was not walking.

DISCUSSION

The dipygus is an extremely rare congenital malformation. Our observation is the 15th case reported in the literature worldwide since the first case published in 1831 by Saint Hilaire and Guerin seen in an 8 year old child. The case we are describing is the second noted in Senegal after the one operated in Belgium and published in 1979 by Braun ($_1$). Other cases were described in Africa: Bankole-Sanni ($_2$) in Ivory Cost published the 10th case in 1996 as Cywes ($_3$) in

South Africa reported in a series of twins two cases of dipygus in 1997.The case before the one we have found was in an American child ([4]).All clinical forms may be encountered, from a third surnumerary arm considered as an incomplete twin to complete caudal duplication associated with genito-urinary and digestive duplications. The search for associated malformations must be systematic in any case of dipygus because this determines the vital and functional outcome.

Plain spine X-rays, ultrasonography and computed tomograghy are of great value. These not only help to clarify the diagnosis and to search for associated malformations but also to guide the surgical management. This surgical management presents various difficulties. The forms of dipygus in which there are one or more surnumerary arms of the pelvis have much easier surgical management. This consists of the resection of the surnumerary leg and pelvis. However the more complicated forms of caudal abnormality are more difficult to manage. The reconstruction of the genito-urinary and gastrointestinal tract is difficult. Massive resections are sometimes necessary with poor end results ([5,6]).

The case we are presenting is a simple form of caudal abnormality with a third leg made of a femur articulating with a proximal left sacro-iliac joint and a distal rudimentary tibia.

This case is comparable to the one reported by Bankole-Sanni et Herman, a part the myelomeningocele encountered in our case. We did not perform a CT scan or MRI as the patient could not afford them. An arteriograph was done that helped the surgical procedure by ligating first the vascular pedicle before the resection of the surnumerary leg. The post operative period was uneventful but the child did not walk up to the age of three probably because of the sacral abnormality.

CORRESPONDANCE

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