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Case Report

The Challenge of Omphalo-ischiopagus Siamese Twins

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Abstract

Introduction: We present the surgical one-stage approach for the separation of a case of severely malformed omphalo-ischiopagus Siamese Twins.

Objective: Treatment is conditioned by what nature presents to us. In this case, the presence of a single penis in those male babies, entailed that one had to become a morphological female, although genetically male.

Material/Methods: There was only a rectal fistula for both, that became the vagina for the "new" girl (to whom testicles were removed). Having 2 bladders but a single Y shaped urethra, the short arm became the new girl's urethra and the long one, terminating in an epispadias (simultaneously repaired), was kept for the boy. The colon was divided in three segments, the distal one for the vagina and the 2 proximal ones being used for a pull through (reconstructing an anus). Shared liver and small bowel were divided, one half attributed to each patient. Sacral osteotomies were required to approximate the pubic bones. Finally the abdominal wall was closed, trying to reconstitute a simulated umbilical scar.

Discussion/Conclusion: Survival depends basically on the extent of the malformation. The Surgery of Siamese Twins undoubtedly needs Team Work, with special relevance for the Anesthesia, aiming to preserve as much as possible normalcy to both twins, particularly concerning their future life.

Keywords: Siamese Twins; Congenital Malformations; Urological Problems; Sexual Differentiation; Rectal and Colonic Malformations; Pelvic Malformations

Introduction and Objectives

The main aim of this paper is to demonstrate the feasibility for reconstruction of an extremely complex congenital malformation and emphasize the mistake of performing an abortion as soon as ultrasound studies reveal a Siamese Tweens pregnancy. It also should serve to demonstrate that the treatment applied is basically conditioned by what nature has provided and requires a wide multidisciplinary competence.

Material and Methods

Male Siamese omphalo-ischiopagus twins, 17 months of age at the time of operation, were treated at the Lisbon Children's hospital D. Estefânia. They had separate hearts, fused livers, common colon opening in the perineum through a single orifice, equidistant from the gluteal folds, 2 bladders (each receiving one ureter from the other twin, a Y shaped urethra with an epispadias meatus (of a single penis), opening near the rectoperineal fistula, with only a

palpable testicle in rudimentary hemi-scrotum, a single umbilical scar.

Having available only one penis for both, the decision was taken to end up with a boy and a girl.

Figure 1: Omphalo-ischiopagus twins and close perineal view.

Surgical technique

After careful clinical and radiological evaluation, under endotracheal anesthesia in a warm operating theater, considering that operation would take many hours (13 and a half in this case).

Considering the possibility of later difficulties for closure of the abdominal wound a previous placement of two 250 ccs expanders were placed in the hipochondrium of both children, but what later proved to have been unnecessary.

Surgery was started with a median transumbilical abdominal incision.

Then followed partition of the liver, the patients having fortunately 2 distinct biliary systems.

The colon was divided in three parts: The distal portion, including the recto-perineal fistula, was used for constructing a neo vagina for the girl, the two proximal halves, with their double vascular supply, were separated to allow, for in each of them, a pull-through operation (after localizing the right place for the new rectal orifice with the help of an electric stimulator (individualizing the external sphincter). Appendectomy of the single appendix was performed.

Figure 2 The bladders were sectioned in two halves till the level of the bladder neck, with the incisions preserving it (so to keep the normal relationship of the ureters) It the girl it was necessary to re-implant an ectopic ureter (not suspected before operation, even with IVP, CT, MRI, Cystography!) The urethra was sectioned at the level of the bifurcate, the short arm being used for the female urethra and the long one for the epispadias male. The epispadias was then corrected with an island flap.

Figure 2: Double small bowel and Pull through.

Figure 3: Bladders receiving ureters from both and pelvic bones and Y shaped urethra.

In the girl the testicles were removed and in the boy an orchidopexy was performed on the right followed by removal of a rudimentary abdominal testicle (placed near the lower pole of the left kidney).

In both a posterior bi-iliac osteotomy was performed to allow for approximation of the pubic rami and closure of the abdominal wound (only then it was realized that the use of expanders was not needed and that their previous placement had been unnecessary (a lesson to be learned).

An onfaloplastia was also performed for aesthetic reasons.

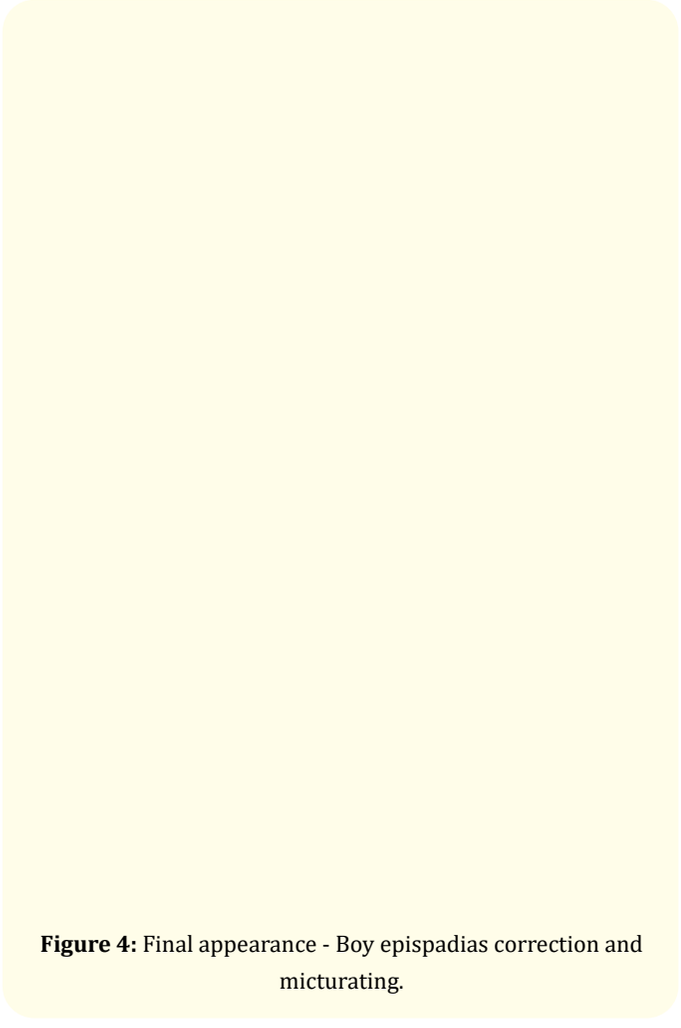

Figure 4: Final appearance - Boy epispadias correction and micturating.

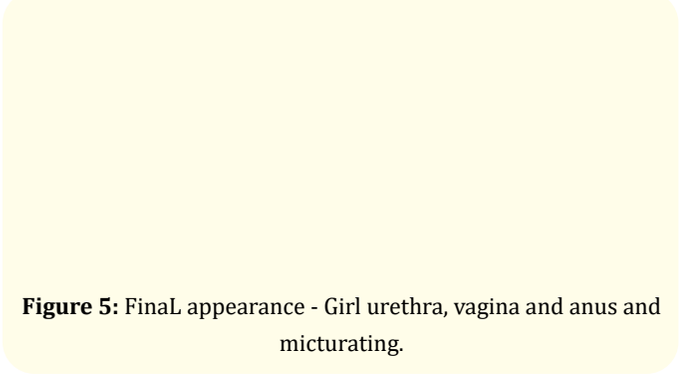

Figure 5: FinaL appearance - Girl urethra, vagina and anus and micturating.

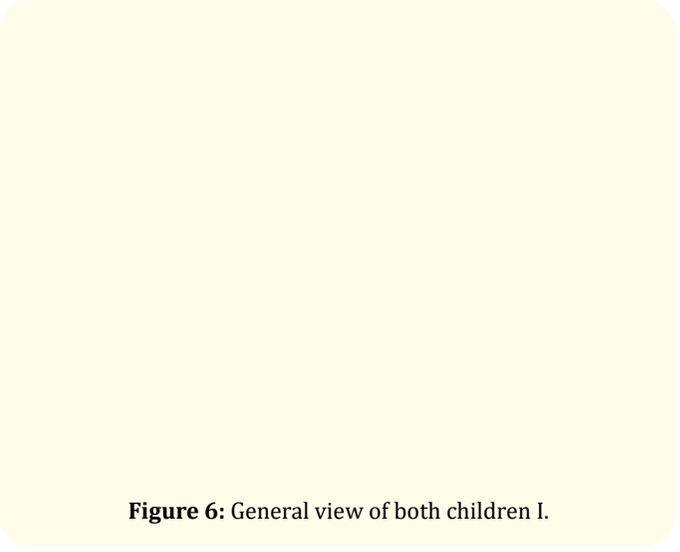

Figure 6: General view of both children I.

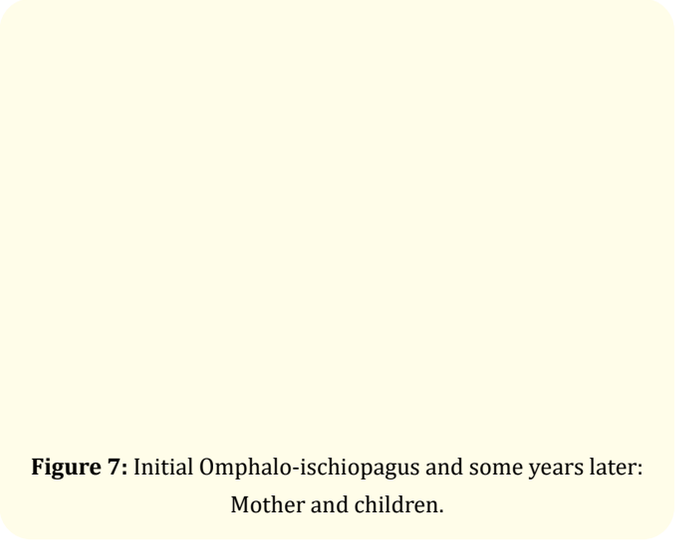

Figure 7: Initial Omphalo-ischiopagus and some years later: Mother and children.

Discussion

We believe that Siamese twins are monozygotic twins in whom separation was not completed and I can not agree with the fusion theory. They may remain joined through any part, with higher or lower intensity, so defining the designations utilized. Their frequency is considered to be 1 to 45.000 to 200.000, accepting that another 40% have probably died in utero and the great majority died soon after birth. The case we present represents probably no more than 2% of all Siamese twins born. Some Siamese Twins that survive may live many years, such as the famous Chang and Eng Bunker (they married two sisters and had 22 children….!.). They originated the name having been born in Thailand (then the Siam Kingdom).

Survival depends basically on the extent of the malformation: nevertheless, nowadays in many places of the so called civilized Countries (where ultrasound is routinely used in pregnancy surveillance), the wrong decision of abortion is taken even in patients that could survive as normal as any other human being.

The more frequent types are the thoracopagus (28%) and thoraco-omphalopagus (18,5%) fortunately the ones with a better prognosis after separation (unless having only a common heart, normally greatly malformed and that is not useful for any of them, who will die usually a few weeks after birth). Surprisingly this anomaly is much more frequent in females than in males, in a proportion of about 3 to 1.

In cases that survive but are not separated, the death of one leads always to the death of the other. Some types like the Parapagus caudalis (with a single lower body and a duplicated upper body) raise the impossible ethical solution of choosing the Right or the Left "head".

It is thought that the first Siamese twins were born in Peru, referenced in the Moche Culture of the III century. 1n 415 Saint Augustine referred to parapagus caudalis siamese twins but the first survivors following surgery seem to have been From Switzerland (Basileia) in 1989 (omphalopagus), followed by Brasil, in 1900 (xiphopagus) and the USA (Minnesota) in 1987 (craniopagus).

We have operated 7 pairs with the survival of 9, the 1st one (Thoraco-omphalopagus) as far as 1978. Then thoraco-omphalopagus (1984), Omphalopagus (1986), Xipophagus (1988) and omphalo-ischiopagus (1999). In this last pair the girl will require periodic vaginal dilatations under general anesthesia and, at puberty, hormonal support with estrogens.

Figure 8 Siamese that survived, operated at Lisbon's Children's Hospital D. Estefânia (172).

Figure 8: Siamese twins that survived after operation at the Children´s Hospital D. Estefânia).

In 1979 2 ischiopagus died of malignant hipertermia (after successful surgical repair, in 1984 2 parapagus (one already death just at birth) and in 1986 1 omphalopagus 2 months old and weighing 2.5 Kg, died 1 month after surgery in an adult's Intensive Care Unit [1-10].

Conclusions

- Siamese twins are independent human beings, with their dignity, intrinsic reality and rights.
- The Surgery of Siamese Twins means is undoubtedly Team Work, with special relevance for the anesthesia, in operations that last many hours. Their therapy requires competent multidisciplinary professionals, but, as with all orchestra's work, there is always a need for a Maestro
- Even with modern technologies the surgical team must be prepared for surprises and for immediate straight forward decisions. Treatment is conditioned for what nature has provided, one aiming to preserve as much as possible normally to both twins, particularly concerning their future life.

Disclaimer

The paper is a single author piece and presents no conflict of interests.

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