

Successful Separation of Conjoined Twins at the Conakry University Hospital Center

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Abstract

Introduction: The birth of conjoined twins is an event extremely rare that offers unique therapeutic challenges and circumstances. We must examine each situation with the many questions that arise and which sometimes require a long reflection. We report a case of separation of pygopages conjoined twins as well as a review of the literature. **Patients and Observations:** A pair of conjoined twins aged 11 days, weighing 3080 g between them, was referred to the neonatology department from the Faranah prefecture, 300 km from Conakry, for care. The mother, a 30-year-old housewife, multipara, eight gestures and nine parities, had not followed any prenatal consultation. The delivery took place at home in a village with the death of the mother immediately postpartum. The clinical and paraclinical investigation had concluded with the diagnosis of Siamese type pygopage. After multidisciplinary consultation, the surgical treatment by separation of the twins was carried out at the age of 50 days with success and preservation of the physiological functions. **Conclusion:** The birth of conjoined twins is extremely rare. Each pair of Siamese is different and the surgical strategy must be adapted according to the shared organs. The perfect multidisciplinary work of the medical staff has been the main contributor to our success.

Keywords

Siamese Twins, Pygopages, Multidisciplinary, Separation, Conakry Hospital and University Center

1. Introduction

The term “Siamese” originates from Chang and Eng Bunker, fused twins from

Siam, Southeast Asia, and joined by the waist. They went to Paris under the Second Empire for surgery. Considered impossible at the time, their separation could not take place [1]. Each case of birth and eventual separation from conjoined twins is unique and extremely rare. Although multiple pregnancies are on the rise worldwide, mainly linked to the older age of future mothers and the use of fertility treatments, the number of joint twin births remains marginal: one per 100,000 births in the world, on average. The majority of conjoined twins born in France are children whose mother comes from a country that does not systematically offer an ultrasound in the first trimester. When it appears that the fetuses are indeed conjoined, the high risk of intrauterine death, birth mortality, complications and serious disorders after birth leads the vast majority of parents to decide to terminate the pregnancy. More than 40% of these children die in utero or at birth and 30% of those who survive birth but die within 24 hours. Since the very first attempt recorded in the tenth century, specialists estimate that until 2015, there have been 250 successful separation procedures worldwide and thousands of refusals to intervene [2].

The surgery to separate Siamese can, depending on the meeting point, be very complex and of very high risk, even fatal depending on the organs that are connected. In addition to purely technical questions, separation often raises serious ethical questions, especially when they share a vital organ: should the good health, or even the life, of one of the twins be sacrificed for the benefit of the other? Advances in imaging that allow a precise vision of all organs in 3 dimensions, as well as the arrival of 3D printers to manufacture exact replicas of the parts to be operated, on have enabled successes that were impossible before [2]. The anesthetic management of conjoined twins has demonstrated the advantages of extensive preoperative investigations and meticulous planning by a multidisciplinary team [2]. Pygopagus twins are joined on the dorsal side of the pelvis sometimes showing sacral abnormalities, a shared anus, and no small bowel abnormalities. They make up about 10% to 20% of all conjoined twins with a female predilection of up to 80% [3]. The results of surgical treatment are often better (80%) [3].

In Guinea, no successful Siamese separation surgery had been performed so far. The first case of the successful separation of conjoined twins recorded in the country was omphalopages, all also boys, who were separated in May 2015 at Necker Enfants Malades Hospital in Paris, France.

So, would separation surgery for all cases of conjoined twins be reserved exclusively for developed countries? It is to answer this question that we present this case of conjoined twins type pigopagus with a review of the literature on the approaches of care.

2. Patients and Observations

A pair of conjoined twins aged 11 days, weighing 3080 g between them, male, was referred to the neonatology department of the Donka national hospital from

the Faranah prefecture located 350 km from Conakry, the capital for support. The mother, a 30-year-old housewife, multipara, eight gestures and nine parities, had not followed any prenatal consultation. The delivery took place at home in a village located 7 km from the capital of the sub-prefecture, 64 km from the prefecture. Immediately after the birth, the parents noticed the anomaly. The mother, who was in poor general condition, died four hours later of hemorrhage. The Siamese were later transported by motorcycle taxi to the prefectural hospital from where they were referred to the neonatology department of the Donka national hospital. After hospitalization, a multidisciplinary opinion was given in particular nutritional, pediatric surgery, resuscitation, neurological and cardiological.

The clinical examination noted in the twins, a good general state, awake, tonic with a good reactivity, the integuments and the conjunctivas normo colored, presence of a thrush, the archaic reflexes are normal, the fontanelles normo tense. They are united by the sacro-coccygeal region in the dorsal position, the deep palpation of which demonstrates a connection between the two coccyxes surrounded by soft tissues. 1); the external genital organs are of the male type, the penis is well developed, the scrotum bifid, the gonads are present at the bottom of the bursae, of normal size and consistency. The abdomen is of normal volume, supple with no palpable mass. The thorax is symmetrical, without morphological abnormality, the lungs are free, the respiratory rate equal to twenty cycles per minute. The spines are separated without visible abnormality; the thoracic and pelvic limbs are without anomaly of axis or structure, nor of apparent malformation. Sensitivity and motor skills are preserved.

The other multidisciplinary opinions did not note any particularity. Heart rate 142 attmet/min, oxygen saturation equal to ninety-seven percent. We retained the diagnosis of Conjoined twins conjoined type pygopage (**Figure 1**).

The X-ray showed a normal skeleton, the spines well separated and connected by the last sacral part at the level of the coccyx (**Figure 2**). The abdominal ultrasound did not note any particularity and the cardiac ultrasound normal except for a lack of use of the color Doppler. The colostogram revealed two colons with completely separated anuses, well opacified and without abnormalities (**Figure 3**).

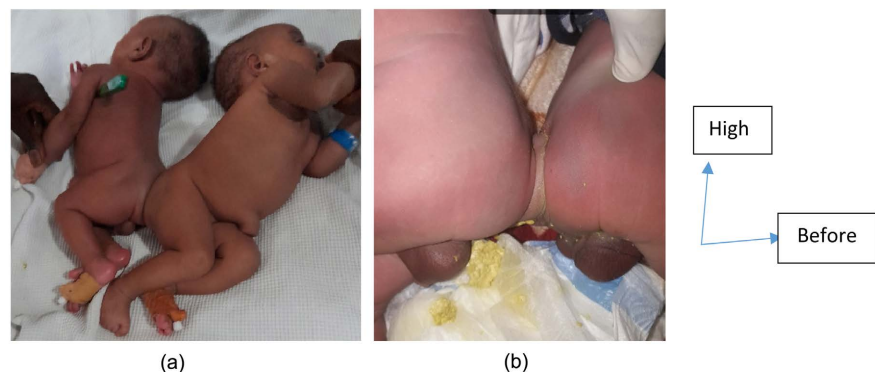


Figure 1. Siamese pygopages in (a); in (b) connecting zone, 2 anal orifices and 2 distinct scrotums present scrotum.



Figure 2. On the left, X-ray of the 2 spines showing the cartilaginous connection between the 2 coccyxes. On the right the colostogram shows the independence of the 2 colonists.

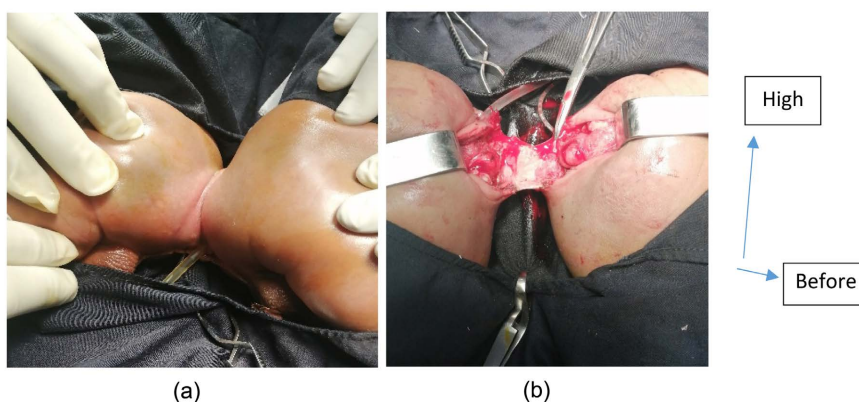


Figure 3. In (a), exposure of the approach, in (b), complete separation of the muscular and rectal structures holding only by the skin.

The biological assessment was unremarkable. They benefited from basic rehydration with Ringer's lactate 80 ml/kg/24hours associated with hypertonic glucose serum at 30%, a diet according to protocol, a mouthwash using sodium bicarbonate and an antifungal made from nystatin syrup 1 pipette 3 times a day. The pre-anaesthetic consultation validated the separation surgery subject to the use of two anesthetic devices.

After three multidisciplinary and interdisciplinary consultation meetings, the indication for a separation was raised. On the eve, the entire system was set up, including 2 anesthetist devices with all the resuscitation equipment and two operating tables. The Twins were taken to the theater under general anesthesia, in right lateral decubitus for the first twin and left for the second. We placed Charrière 6 urethral and rectal probes. At the first stage, we made a midline incision in the furrow located at the junction of the two buttocks from top to bottom, involving the skin and the subcutaneous cell tissue with an ordinary scalpel and then hemostasis by electrocoagulation. Median opening at the junction between the posterior fibers of the levator ani muscles then progressive dissection. Indi-

vidualization by dissection of the posterior wall of the rectums was joined. Highlight then median section with an electric bistoury of the fusion zone of the coccyx. Identification and mucious dissection of the cleavage zone between the two posterior walls of the rectum allow their separation. Median opening at the junction between the fibers of the posterior part and the fibers of the external sphincter, and then last cutaneous opening in depth allows the complete separation of the twins (**Figure 4**).

In the second stage, the team of four surgeons was immediately divided and the twins were repositioned in ventral decubitus, block under the pelvis on separate operating tables. Repair of the fibers of the levator ani muscles and the external sphincter by anchoring separate points of vicryl 4/0. Suture of the subskin and the skin, separate 4/0 vicryl stitches. In the end, obtain a well-contractile anus (**Figure 4**). The duration of the intervention was 2 hours 30 minutes.

The twins woke up fine after 30 minutes in the recovery room. The multidisciplinary follow-up was continued and at the last follow-up of 8 months the twins are doing well with normal continence.

3. Discussion

The cases of Siamese babies are a rare event that the majority of therapists will not encounter during their professional career [4]. The exact number of surviving cases until complete separation is not really known [2]. Conjoined twins are monozygotic, mono amniotic and mono chorionic twins. Two conflicting theories exist to explain the origins of conjoined twins. The first, the most accepted, fission, in which an incomplete division on the thirteenth day of gestation produces identical twins with common anatomical structures. The second, the fusion in which two completely separated fertilized egg embryos interact between the thirteenth and seventeenth day of gestation. More recent studies reject these theories for dorsally united twins and suggest a secondary fusion of originally separated twins [2]. Spencer in 2003 stated that conjoined twinning originates in the first week after fertilization [4].

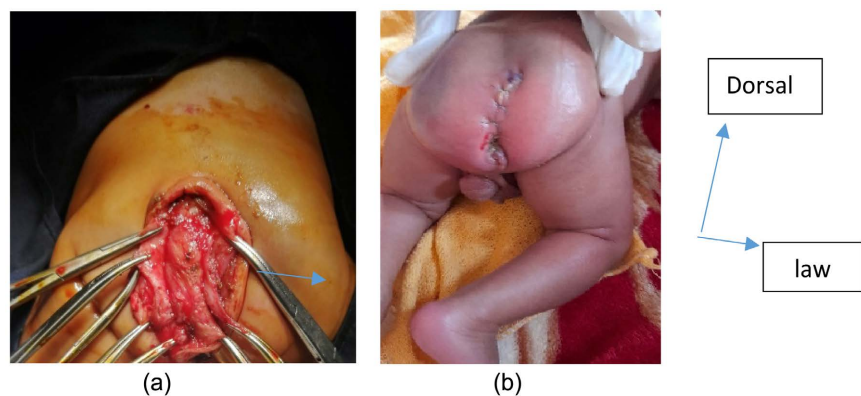


Figure 4. View in (a) of the identification of the different muscular planes before mooring to the rectum. In (b) after closing.

Each set of twins poses unique anatomical challenges and general principles should be adapted to each circumstance [3]. Caudal variants (11%) include ischiopagus [2]. Pygopagus twins are united at the sacrum, coccyx and perineum. The union often involves the dural sheath and the terminal part of the spinal cord. Structures arise from secondary neurulation (conus medullaris and filum terminale), arising after closure of the caudal neuropore and common in pygopagus. The degree of dural conjunction and spinal and perineal cord, genitourinary and sacrococcygeal morphology are the most important considerations for a possible separation [2]. In our case, the union involved only the terminal part of the spinal cord and the posterior perineum with some muscle fibers of the pelvic diaphragm. The multidisciplinary team approach plays an important role in obtaining a satisfactory final surgical result [5]. The reconstruction of the rectal anatomy is based on the usual imperforation. Surgical principles and continence can be a postoperative issue. Magnetic resonance imaging (MRI) can help define the pelvic musculature and help plan the separation. Evaluation of the genitourinary, anorectal, skeletal and neurological systems and the search for other abnormalities will help determine the extent of morbidity after separation [3]. Given the limitation of means in our environment, the anatomical site of the union considered favorable and the non-existence of associated congenital anomalies proven by the multidisciplinary consultation, we carried out ultrasounds and x-rays of the skeleton with a colostogram. This assessment allowed us to organise, plan and anticipate reconstruction needs before the separation. The intervention, always at high risk, was planned gesture by gesture for the two teams who separated as soon as the bond between the two children was broken.

Anesthetic difficulties are related to anatomical barriers and the degree of cross-circulation between the twins. In this case ethical problems arise if the separation involves an unequal sharing of limbs and organs or when the separation results in the death of one of the twins [2]. Our team did not encounter this kind of difficulty because the reconstruction of the rectal anatomy was based on the usual imperforation that we regularly practice and the challenge was the surgical principles and continence, the non-respect of which can be a postoperative problem. The anesthetic challenge was to simultaneously intubate and ventilate the twins who were facing backwards [3]. Siamese require a nutritional assessment. Adequate nutrition therefore remains imperative to support growth and development during the neonatal period [6]. The staff of the nutrition service in collaboration with that of the neonatology unit had invested in the pre and postoperative period to feed the twins correctly. This state of affairs contributed to the planning and success of the work of the operating room team.

Regarding the age of the intervention of the Siamese, the majority agrees between 3 and 6 months after birth [7]. Emergency separation may be necessary in the event of damage to the connecting bridge (omphalopagus), or if one of the twins threatens the life of the other (complex congenital heart disease, cardiomyopathy, sepsis), when the deterioration of both twins occurs due to hemodynamic and respiratory compromise (typically thoracopagus) or when the con-

dition of one of the twins is incompatible with life (anencephalic) [8]. In our case, the separation was considered early at the age of one month three weeks because of the multidisciplinary collaboration, the simplicity of the anatomical site of connection, the absence of associated malformation, lack of sufficient financial means and fear of deterioration in their condition. Measures such as prenatal consultations, regular follow-up of pregnant women in order to carry out early detection of pregnant spouses and to prepare early for their care, must be taken into account. The preparation and management of the birth by the obstetrics and neonatology teams is already a real challenge. The pregnancy will be monitored every fortnight and the possible interventions evaluated very quickly in order to prepare them as soon as possible [7].

In the study by Fallon SC *et al.* [3], all cases reported up to 1990, with 167 separations listed, overall survival was 64%, with better results in pyopages and ischiopages up to 80%. He also reported that the male pygopage conjoined conjoins were more susceptible to mortality but those that survived had more favorable rectal anatomy. This favorable anatomy was found in our case. Contrary to their observation, the 2 cases of Siamese recorded in our country were all male and survived. Suggestions for avoiding problems in the future should relate to the constitution not multidisciplinary teams dedicated to the care of Siamese, training focused on the management of Siamese and the creation not association of joint surrogate mothers.

4. Conclusion

The surgical separation of conjoined twins represents one of the most unique operative procedures. Surgical principles such as a thorough analysis of the understanding of embryology and anatomy, thorough preoperative planning, the involvement of the entire multidisciplinary team and the surgical approach form the basis of a successful operation.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

References

- [1] Heuer, G.G., Madsen, P.J., Flanders, T.M., Kennedy, B.C., Storm, P.B. and Taylor, J.A. (2019) Separation of Craniopagus Twins by a Multidisciplinary Team. *The New England Journal of Medicine*, **80**, 358-364. <https://doi.org/10.1056/NEJMoa1805132>
- [2] Frawley, G. (2020) Conjoined Twins in 2020—State of the Art and Future Directions. *Current Opinion in Anesthesiology*, **33**, 381-387. <https://doi.org/10.1097/ACO.0000000000000864>
- [3] Fallon, S.C. and Olutoye, O.O. (2018) The Surgical Principles of Conjoined Twin Separation Seminars in Perinatology. *Seminars in Perinatology*, **42**, 386-392. <https://doi.org/10.1053/j.semperi.2018.07.013>
- [4] Boer, L.L., Schepens-Franke, A.N. and Oostra, R.J. (2019) Two is a Crowd: On the

Enigmatic Etiopathogenesis of Conjoined Twinning. *Clinical Anatomy*, **32**, 722-741. <https://doi.org/10.1002/ca.23387>

- [5] Wu, S., Guo, K., Xiao, P., Yang, J. and Sun, J. (2018) Body Wall Reconstruction For Conjoined Twins Our Experience And Lessons Learned. *Annals of Plastic Surgery*, **81**, S66-S70. <https://doi.org/10.1097/SAP.0000000000001609>
- [6] Bergner, E.M., Gollins, L., Massieu, L.A., Hurst, N. and Hair, A.B. (2018) Nutritional Considerations in the Care of Conjoined Twins. *Seminars in Perinatology*, **42**, 355-360. <https://doi.org/10.1053/j.semperi.2018.07.018>
- [7] <https://sante.lefigaro.fr/article/des-soeurs-siamoises-reliees-par-la-tete-ont-ete-separes-avec-succes/>
- [8] Cummings, B.M. and Paris, J.J. (2019) Conjoined Twins Separation Leading to the Death of One Twin: An Expanded Ethical Analysis of Issues Facing the ICU Team. *Journal of Intensive Care Medicine*, **34**, 81-84. <https://doi.org/10.1177/0885066618791953>