

Successful Separation of Siamese Twins at Conakry University Hospital

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ABSTRACT

Introduction: The birth of conjoined twins is an event extremely rare which offers unique therapeutic challenges and circumstances. Each situation must be examined with the many questions that arise and which sometimes require a long reflection. We report a case of separation of pygopage twins as well as a review of the literature.

Patients and Observation: A pair of conjoined twins of 11 days of life, weighing 3080g between them, was referred to the neonatal service from the Faranah prefecture, 300 km from Conakry, for treatment. The mother, a 30-year-old housewife, multiparous, eight gestures and nine parities, had not followed any prenatal consultation. The birth took place at home in a village with the death of the mother in the immediate postpartum period. The clinical and para-clinical investigation had concluded with the diagnosis of Siamese pygopage type. After a multidisciplinary consultation, the surgical treatment by separation of the twins was carried out at the age of 50 days with success and conservation of 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 nursing staff has been the main contribution to our success.

Keywords

Siamese twins, Pygopages, Multidisciplinarity, Separation, Conakry hospital and university center.

Introduction

The term "Siamese" originates from Chang and Eng Bunker, merged twins from Siam, South-East Asia, united by the size. They went to Paris under the Second Empire for surgery. Deemed impossible at the time, their separation could not take place [1]. Each case of birth and eventual separation of conjoined twins is unique and extremely rare. While multiple pregnancies are on the increase worldwide, mainly due to the older age of future mothers and the use of fertility support treatments, the number of joint twin births remains marginal: one per 100,000 births around the world, on average. The majority of conjoined twins who are born in France are children whose mother arrives from a country which 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, death at birth, complications and serious disorders after birth leads the vast majority of parents to decide to terminate the pregnancy. Over 40% of these children die in utero or at birth and 30% of those who survive birth die within 24 hours. Since the very first attempt recorded in the 10th century, specialists estimate that there were until 2015, 250 successful separation interventions in the world and thousands of refusals to intervene [2].

The surgery to separate Siamese can, depending on the meeting point, be very complex and very high risk, or even fatal depending on the organs that are connected. In addition to purely technical questions, separation often poses 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 which provide a precise view of all

organs in 3 dimensions, as well as the arrival of 3D printers to make exact replicas of the parts to be operated on have enabled successes previously impossible [2]. The anesthetic management of conjoined twins has demonstrated the advantages of extensive preoperative investigations and careful planning in a multidisciplinary team [2]. Pygopagus twins are joined on the dorsal side of the pelvis sometimes exhibiting sacral abnormalities, a shared anus and no small bowel abnormalities. They represent around 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 successful twinning separation case recorded in the country was omphalopagi, also male, who were separated in May 2015 at Necker Enfants Malades Hospital in Paris, France.

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

Patients and Observation

A pair of conjoined twins aged 11 days of life, weighing 3080g between them, male, was referred to the neonatology service 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, multiparous, eight gestures and nine parities, had not followed any prenatal consultation. The birth took place at home in a village located 7 km from the capital of the sub-prefecture, 64 km from the prefecture. Immediately after birth, the parents noticed the anomaly. The mother, who was in poor general condition, died four hours later, from hemorrhage. The Siamese were subsequently transported by motorbike 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 on nutrition, pediatric surgery, intensive care, neurology and cardiology.

The clinical examination noted in the twins, a good general state, awake, toned with a good reactivity, the integuments and the conjunctiva normo colored, presence of a thrush, the archaic reflexes are normal, the fontanelles normo tense. They are united by the sacrococcygeal region in the dorsal position whose deep palpation shows a connection between the two-coccyx surrounded by soft parts 1); the external genitalia are male, the penis is well developed, the scrotum bifid, the gonads are present at the bottom of the bursa, of normal size and consistency. The abdomen is of normal size, flexible without a palpated mass. The thorax is symmetrical, without morphological anomaly, the lungs are free FR = 40 cycles / m. The spines are separated without any visible abnormality; the thoracic and pelvic limbs are free from axis or structural abnormalities, nor any apparent malformation; sensitivity and motor skills are preserved.

The other multidisciplinary opinions did not note any particularity. Heart rate 142attemet / min, O2 saturation = 97%. We retained the diagnosis of conjoined Siamese twins of the pygopage type.



Figure 1: Siamese pygopages in A; in B connection area, 2 anal openings and 2 separate scrotum with scrotum.

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 subject to a failure to use the color Doppler. The colostogram revealed two colonists with completely separate anuses, well opacified and without anomalies (Figure 3).

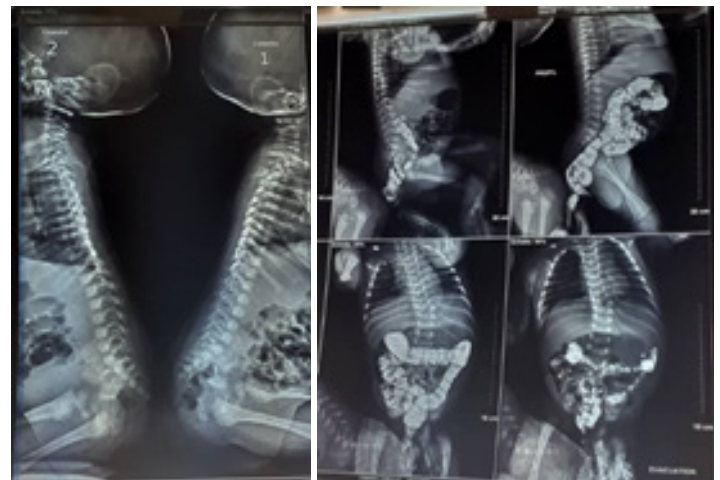


Figure 2: Left, x-ray of the 2 spines showing the connection between the 2 coccyxes. On the right the colostogram showing the independence of the 2 colonists.

The biological assessment was unremarkable. They benefited from basic rehydration with Ringer lactate 80ml / kg / 24h associated with hypertonic glucose serum at 30%, a diet according to protocol, a mouthwash using sodium bicarbonate and nystatin syrup 1 pipette 3 times a day. The pre-anesthetic 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. The day before, all the device was put in place including 2 anesthetic machines with all the resuscitation equipment and two operating

tables. The Twins were taken in the operating room under general anesthesia, in the right lateral decubitus for the first twin and left for the second. We placed Charrière 6 urethral and rectal stent probes. First, we made a midline incision in a groove located at the junction of the two buttocks from top to bottom, involving the skin and the subcutaneous cellular tissue with an ordinary scalpel, then hemostasis by electrocoagulation. Median opening at the junction between the posterior fibers of the levator ani muscles followed by progressive dissection. Individualization by dissection of the posterior wall of the rectums which were contiguous. Demonstration then mid-section with an electric scalpel of the coccyx fusion zone. Identification and mucinous dissection of the cleavage zone between the two posterior walls of the rectums allowing their separation. Median opening at the junction between the fibers of the posterior part and the fibers of the external sphincter, then the last deep cutaneous opening allowing the complete separation of the twins (Figures 4).

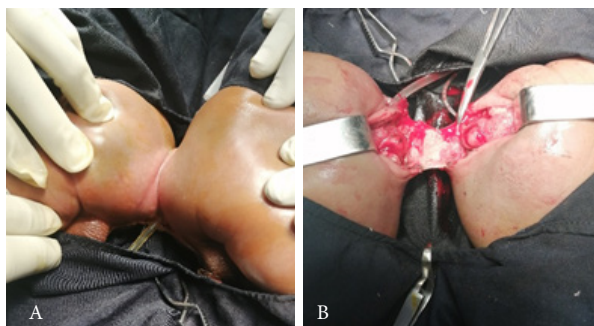


Figure 3: in A, exposure of the first approach, in B, complete separation of the muscular and rectal structures holding only by the skin.

In the second step, the team of four surgeons immediately split up and the twins were repositioned in prone position, chopping blocks 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 4/0 vicryl. Suture of the sub-skin and of the skin, separated points of vicryl 4 / 0. At the end, obtaining a well contractile anus (Figure 6). The duration of the operation was 2 hours 30 minutes.

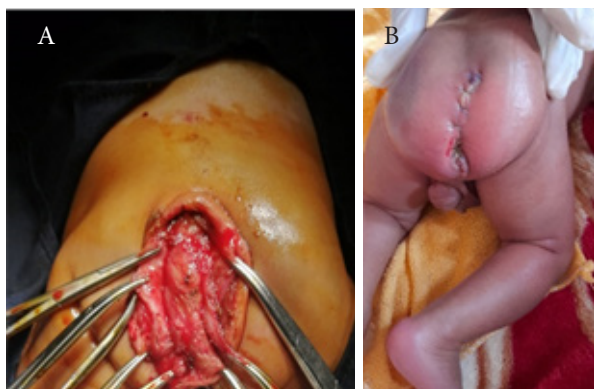


Figure 4: A view of the location of the different muscular planes before anchoring to the rectum. In B after closing.

The twins woke up well 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.

Discussion

Siamese babies cases are a rare event that the majority of therapists will not encounter in their professional career [4]. The exact number of cases surviving until complete separation is not fully known [2]. Conjoint twins are monozygotic, mono amniotic and mono chorionic twins. Two contradictory theories exist to explain the origins of the Siamese twins. The first, fission, the most accepted, in which incomplete division on the 13th day of gestation produces identical twins with common anatomical structures. The second, the fusion in which two completely separated embryos of fertilized eggs interact between the 13th and 17th days of gestation. More recent studies reject these theories for dorsally united twins and suggest secondary fusion of initially separated twins [2]. Spencer in 2003 stated that joint twinning originates in the first week after fertilization [4].

Each group of twins poses unique anatomical challenges and the general principles must be adapted to each circumstance [3]. Caudal variants (11%) include ischiopagus [2]. The Pygopagus twins are united at the level of the sacrum, coccyx and perineum. The union often involves the dural sheath and the terminal part of the spinal cord. Structures originating from secondary neurulation (medullary cone and filum terminale), occurring after closure of the caudal neuropore and common in pygopages. The degree of dural and spinal and perineal, genitourinary, and sacrococcygeal marrow conjunction are the most important considerations for 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 from the pelvic diaphragm. The multidisciplinary team approach has an important role in obtaining a satisfactory final surgical result [5]. Reconstruction of the rectal anatomy is based on the usual imperforation. Surgical principles and continence can be a postoperative problem. Magnetic resonance imaging (MRI) can help define the pelvic musculature and help plan separation. Assessment of the genitourinary, anorectal, skeletal and neurological system and the search for other abnormalities will help determine the extent of morbidity after separation [3]. Given the limited resources in our environment, the anatomical site of the union deemed favorable and the inexistence of associated congenital anomalies proven by the multidisciplinary consultation, we performed ultrasounds and x-rays of the skeleton with a colostogram. This assessment enabled us to organize, plan and anticipate reconstruction needs before separation. The intervention, still 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.

The anesthetic difficulties are related to the anatomical barriers and the degree of cross circulation between the twins. In this case ethical problems arise if the separation involves an unequal division 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 has been based on the usual imperforation that we practice regularly and the challenge has been the surgical principles and continence whose non-observance can be a postoperative problem. The anesthetic challenge was to simultaneously intubate and ventilate the twins with their backs [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 department in collaboration with that of the neonatal unit had invested in the pre and postoperative period to properly feed the twins. This state of affairs contributed to the planning and success of the work of the operating room team.

Regarding the age of intervention of Siamese, the majority agrees between 3 and 6 months after birth [Figaro]. Emergency separation may be necessary in case 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 condition of one of the twins is incompatible with life (anencephalic) [7]. In our case, separation was early considered at the age of 1 month 3 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 of their condition. 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 pyopagus and ischiopage twins up to 80 %. He also reported that Siamese pygopage-type males were more susceptible to mortality but those who 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.

Conclusion

Surgical separation of Siamese twins is one of the most unique.

Operating 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.

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