



活水堂

半畝方塘一鑑開 天光雲影共徘徊
問渠那得清如許 爲有源頭活水來

— 宋·朱熹 —

醫學浩瀚，教學研究傳故啓新、開發心靈，
只要智慧活水源源不絕，就能川流大海，成就多元浩瀚之美。
活水堂，傳智啓慧，爲廣博醫海不斷引入源泉活水。

巧手分割脊椎 完美神經重建

有「神經建築師」美譽的美國羅格斯大學教授楊詠威 (Wise Young)，二〇一一年九月應花蓮慈濟醫學中心邀請參加第二屆幹細胞及再生醫療國際研討會時，盛讚花蓮慈濟醫院分割臀部相連的連體嬰玫瑰姊妹「非常不簡單」，因為玫瑰姊妹不只臀部相連，而是脊椎骨尾端相連，是非常困難的術式。他說：「這應該說是脊椎骨相連的連體嬰分割才是！尤其共用生殖與排尿口，困難度更高。」

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Successful separation of the conjoined thecal sac with an epidermal cyst in pygopagus twins

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Abstract Pygopagus conjoined twins are very rare, and half of all pygopagus twins have other anomalies. This report describes the successful surgical separation of pygopagus twins who had a conjoined thecal sac and an epidermal cyst. Metabolic preoperative planning, development of a vascular anastomosis, multistage teamwork, and a staged surgery contributed to a successful outcome. © 2011 Elsevier Inc. All rights reserved.

Pygopagus twins, those joined at the rump, are classified as symmetrical conjoined twins [1]. Pygopagus twins are exceedingly rare, accounting for only 1% to 15% of all conjoined twins, yielding an incidence of approximately 1 in 1,000,000 live births. Pygopagus conjoined twins commonly share the gluteal region, terminal spine, and gastrointestinal, urogenital, and reproductive systems to various degrees. Half of all pygopagus twins have anomalies unrelated to the classic fused organs, including a high incidence of vertebral anomalies [2,3]. This report describes the neurosurgical management of pygopagus twins with spinal anomalies and an epidermal cyst.

1. Case report

Female pygopagus conjoined twins were delivered by cesarean delivery at 39 weeks of gestation in the Philippines. They were transferred to the Buddhist Tzu Chi General Hospital in Taiwan at 5 months of age (Fig. 1). The twins shared a common urogenital sinus with separate vaginal and urethral openings and one anus with low fixation of the rectum. Both twins had good movement of the lower limbs and were each capable of sitting and defecating. Magnetic resonance imaging (MRI) revealed a single dural sac between 2 separate sacral bones (Fig. 2). Based on the amniotic computed tomographic and MRI images, a model of the twins was developed and used for surgical planning and simulation.

To minimize adequate skin flaps to cover the underlying tissue and organs after their separation, the twins underwent staged operations. First, tissue expansion was implanted in

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Fig. 1 Photograph of the pygopagus twins joined back to back. The twins are shown from the back, highlighting their conjoined anatomy.

Fig. 2 Three-dimensional computerized tomographic (CT) scan of the twins' lower back, showing the shared sacral region and the conjoined thecal sac.

2. Discussion

Pygopagus conjoined twins are challenging to treat. The twins resulting on the torso described above, among the challenges they faced, created a novel based on the ongoing work for surgical planning and simulation. The successful separation of these twins also relied on the multidisciplinary team, including expertise and timing of the staged operations. Intraoperative separation necessitated careful blood control, with monitoring rates as high as 300 mL. The physical condition, severe organ anomalies, prolonged surgery times, and significant blood loss, and appropriate transfusion, are common to these patients undergoing this surgery. It is difficult to plan of conjoined conjoined twins separated because of the low survival rate (70% reported at 10 to 15 months of age [1]).

First, due to the physical and psychological condition, separating pygopagus twins using a staged approach may be the best option. First, the physical and psychological condition of the twins should be assessed, and an attention to the conjoined thecal sac was identified and assessed. In the authors' experience, conjoined twins are conjoined twins do not appear to have any previously described. These cases could also have been treated as conjoined twins. These cases could also have been treated as conjoined twins. These cases could also have been treated as conjoined twins.

Separation of a conjoined thecal sac

and divided the neural elements before the bowel was opened to abrogate concerns regarding a contaminated surgical field [1]. The surgical team strived to approximate neural elements as equally as possible to avoid damaging either twin. The option of not sectioning roots between the twins did not exist, as intraoperative physiologic monitoring was precluded to be of little help [1] and was, therefore, not used. The twins had no neurologic deficits after the separation, supporting this decision. Next, primary closure of the dura in these pygopagus conjoined twins was achieved without a graft of patch, both of which have previously been described [7,8]. Despite the fact that CSF leakage in pygopagus twins surviving separation with union of the spinal cord is as high as 37.5% [7] and given that postoperative CSF leakage can necessitate a second surgery [9,10], CSF leakage did not occur in this case. Sacral drains were also avoided to minimize the risk of a CSF leak [11]. Instead, a careful, lightweight dural closure and meticulous myofascial sutures were chosen.

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References

- Agulir E, Ashkenazi J, Buzman W, et al. Conjoined twins in the Philippines: separation of single location. *Pediatr Surg Int* 2008;23:775-80.
- Agulir E, Kelly AM. Experience in the management of conjoined twins. *Br J Surg* 2002;89:1100-02.
- Blackley AD, Gattal P, Walsh R, et al. Management of pygopagus conjoined twins. *Childs Nerv Syst* 2008;24:243-6.
- Shikata Y, Yonemitsu T, Nakano T, et al. Intraoperative epidural and dural cysts. Surgical results of seven cases. *Arch Orthop Trauma Surg* 2009;133:101-4.
- Lee SH, Hwang J, Bock JA, et al. Intraoperative epidural and dural cysts: report of 18 cases and proposal of the treatment. *Pediatr Neurosurg* 2001;35:181-7.
- Frappaz G, Miller A, Roth H. Spinal cord involvement in pygopagus conjoined twins: case report and review of the literature. *Childs Nerv Syst* 2003;19:183-7.
- Mann H, Swisher A, Anshelina A. Anesthetic separation of pygopagus conjoined twins. *J Pediatr Surg* 2004;39:1584-7.
- Geary ME. Pygopagus conjoined twins. *Br Med J* 1962;2:648-71.
- Yoshida TP. Neurologic consequences in a pygopagus conjoined twin. *J Pediatr Surg* 1982;17:551-3.

玫瑰姊妹分割經過的論文，獲得國際肯定，被刊登在《兒童外科期刊》(Journal of Pediatric Surgery) 上。

連體嬰為罕見病例，臀部背側相連連體嬰更為罕見中的罕見病例，多半合併有其他先天異常及中樞神經發育不足，而有類似腦性麻痺之後遺症。菲律賓的玫瑰姊妹經由慈濟醫院醫護團隊集思廣益、分工合作，膽大心細規劃與執行分階段手術，已成功分割連體嬰。其中神經外科團隊，擔負起針對玫瑰姊妹身體最深部——相連脊柱腔及中樞神經髓的畫龍點睛分割部分。

分割脊柱腔及神經髓 國際罕見大師肯定

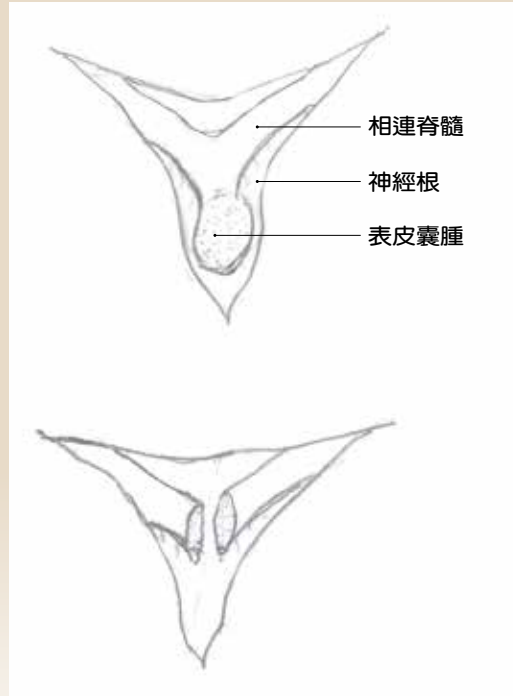
我們將這次的分割所經過的歷次手術模擬會議內容及相關神經手術細節作探討後，很榮幸先在二〇一〇年底的國內神經外科學會年會與國內先進和同儕分享這難得的經驗，他們當下感動讚賞我們濟貧救苦的愛心、慎密的計畫與團隊的執行力。其後經由

科學論文格式的撰寫整理後，今年九月更獲得國際期刊《兒童外科期刊》(Journal of Pediatric Surgery) 的肯定，而以題目——「在臀部相連連體嬰，成功分割脊柱腔及神經髓相連及併有神經髓上皮囊腫」(Successful separation of the conjoined thecal sac with an epidermal cyst in pygopagus twins) 刊載。我們團隊的成果，也在今年九月「第二屆幹細胞及再生醫學國際研討會中」受到國際脊椎損傷治療權威楊詠威 (Dr. Wise Young) 博士的讚揚與鼓勵。

臀部相連連體嬰，佔所有連體嬰之百分之十到十八，發生率為近一百萬活產中有一例。通常他們會共有臀部、尾端脊椎及腸胃、泌尿生殖系統的相連。玫瑰姊妹不同於以往的案例，除脊柱腔及神經髓相連異常外合併有神經髓上皮囊腫，此種情形未出現在以往的文獻中。去年玫瑰姊妹在五個月



哈鐵木爾醫師在一般外科團隊完成分割後，接手分割與修復脊椎的神經。



原本玫瑰姊妹脊髓和神經根相連，並有罕見的表皮囊腫，經過神經外科切除表皮囊腫後，也完成脊髓與神經的分割。繪圖 / 哈鐵木爾

大時，由菲律賓慈濟分會安排轉診至花蓮慈院，由小兒外科彭海祁主任親自至菲律賓診察玫瑰姊妹，並帶回相關資料助益團隊規劃後續來花蓮治療的分工與步驟。入院由小兒科朱家祥主任收治，影像檢查在麻醉科的細心安排全身麻醉下，醫學影像部完成了電腦斷層與核磁共振檢查，其後根據此影像做出 3D 立體模型，供大家反覆模擬，訂定與修正手術計畫。當時在院長領軍下，相關科系主管與同仁參與多次的籌備會，神經醫學科學中心也另外召開會議討論，回顧相關文獻，制定了脊柱腔及神經髓相連分割之方針。

為確保分割後的器官組織有足夠的皮瓣包覆，第一階段先由整形外科團隊置放「組織擴張器」於玫瑰姊妹之

大腿外側，在此次麻醉中，同時執行膀胱鏡、直腸鏡、陰道鏡來探查泌尿生殖和腸胃系統，並開乙狀結腸造口，以避免未來分割處的傷口受到糞便污染。

細膩分配層層分割 保留最佳神經功能

第二階段在玫瑰姊妹七月大時執行，先將玫瑰姊妹半仰臥，我們由小兒外科與一般外科團隊分割生殖器部分，由整形外科團隊移除組織擴張器；之後將她們俯臥，分割至脊椎時，由神經外科團隊接手。我們先打開脊柱腔外露出相連的神經髓，將相連的神經髓硬脊膜切成「Y」字型打開後，將神經髓尾部相連處的神經上皮囊腫切除，使相連的神經髓由「Y」字型變為「V」



二〇一〇年六月分割手術順利成功，帶給菲律賓這對玫瑰姊妹各自獨立的未來。攝影／彭薇勻

字型。此時便有多餘的神經髓硬脊膜形成。再將相連的神經髓由「V」字型分割為「II」型後，將多餘的神經髓硬脊膜上翻與分割的邊緣緊密縫合而完成脊柱腔及神經髓分割。其後由小兒外科與一般外科團隊分割直腸，再由整形外科團隊縫合皮瓣保護體內組織結構。

第三階段在玫瑰兩姊妹九個月大時，將乙狀結腸造口關閉，兩姊妹此時已經分別生活成長兩個月。整體手術後狀況良好，未有任何脊髓液滲漏與神經功能惡化之後遺症。

成功分割連體嬰，有賴於各專業團隊明瞭如何克服挑戰與困難，依影像製成模型來反覆模擬演練，配合藥物與營養支持，與分階段膽大心細確實執行手術，在過去的世界文獻紀錄，



玫瑰姊妹在分割成功一年後，於慈濟醫療二十五周年時與二〇〇三年分割成功的慈恩慈愛一起回到花蓮慈院。圖為雙胞胎們與小兒科團隊合影，前排左起為鄭雅君護理長、小兒部主任鄭敬楓。

緊急分割連體嬰有高達百分之八十的死亡率，而國外專家建議在六至九個月大時分割連體嬰有七成以上之存活成功率。我們團隊汲取世界相關團隊之智慧，於玫瑰姊妹七個月大時完成分割。在神經外科方面，成功分割的關鍵：一、分割神經部分先於腸道，可避免手術之感染。二、儘可能平均分割神經組織，可保有未來兩個個體最大的神經功能。我們非常感恩上人與基金會的支持，院方的統籌，各科室的專業分工，與志工們的呵護關懷，看到玫瑰姊妹分割後歡喜自在，活動自如回到我們慈濟人的娘家——精舍，一切的努力都是值得，真的很感恩有這個機緣發揮我們的專業，陪伴玫瑰姊妹的成長。🍀