The Pygopagus Conjoined Twins: Separation and Perfect Nerve Systems Rebuilt

Professor Wise Young of Rutger University in the US, who is known as the "Nerve Architect", commended Hualien Tzu Chi Medical Center in separating the twin Rose Sisters while attending the second International Conference of Stem Cells and Regenerative Medicine on Sept. 9, 2011. He thought that the complexity of this surgery "should be very difficult," because the twins are conjoined not only the bottoms but also the end of their spines, especially that they shared urogential and reproduction parts, "which made the surgery even more difficult."

Written by Yu-Cheng Chou and Tomor Harnod, neurosurgeon, Hualien Tzu Chi Medical Center

The thesis on separating the Rose Sister, received international recognition, and was published in the "Journal of Pediatric Surgery".

Conjoined twins are rare cases, but pygopagus conjoined twins are even rarer. They often have other congenital anomalies and underdeveloped central nerve, which lead to prognostic symptom similar to cerebral palsy.

The twins nicknamed “Rose Sisters” from Philippine has been successfully separated by Hualien Tzu Chi medical team. Of which, the neurosurgery team was the key part responsible for the most difficult part of their body – conjoined thecal sac and central nerve.

After we investigated the details of the imitating operation meeting contents and related nerve surge details, we shared our experience with professionals.
In 2010, when the Rose Sisters were about five months old, Tzu Chi’s Philippine Chapter arranged them to transfer to our hospital. Director Hai-Chi Peng of the Pediatric Surgery Dept. went to Philippine to examine the Rose Sisters and brought back related information for further evaluation. Then, they were admitted to our hospital and Dr. Chia-Hsiang Chu as their attending Pediatrician. The anesthesiologists carefully arranged the imaging examination so that we completed CT and MRI. Then, the 3D model was made according to the digital exams. This allowed us to repeat simulation, setup and remodel the operation plan.

To ensure there’d be enough tissue covering organs after the separation, the first step is to have the plastic surgery team placing a “tissue expander” at the Rose Sisters’ lateral thigh. During this step, we also performed cystoscopy, proctoscopy, and colposcopy to check the genitourinary and the gastrointestinal systems. We also performed the parallel sigmoid colostomy to avoid future fecal contamination.

**Delicately Assigned Layers to Retain the Best Nerve Function**

The second stage was performed when the Rose Sisters were seven months old. We first placed them facing up. The pediatric surgery and general surgery teams first separating their reproduction part. The plastic surgery team removed the tissue expander. Then we turned them facing down. We, the neurosurgery team, took over separating the spine. We first opened the exposed marrow of the conjoined spinal cavity. We cut the associated nerve marrow into “Y” shape, and

Pygopagus twins are about 10 to 18 percent of all conjoined twins. The chance having pygopagus twins is nearly one out of a million. They usually share the hips, the end of the spine, stomach, and urine and reproduction parts. The Rose Sisters is different from previous cases. Besides the conjoined thecal sac they had an epidermal cyst. This never happened in history.
The separation surgery in June, 2010 was successful. It gave Rose Sisters of Philippine their own independent future.

After successful separation, Rose Sisters returned to Hualien Tzu Chi Medical Center with Tzu-en and Tzu-ai, conjoined twins separated successfully in 2003, for the 25th anniversary of Tzu Chi Medical Mission. They visit the pediatric team to say thank you.

cut off the epidermal cyst which was connecting to the end of the nerve marrow. This made the nerve marrow changed from “Y” shape to “V” shape. By then, there were excess neural myeloid dural formed. We separate the nerve marrow from “V” shape to “II” shape. Then, we turned the excess nerve myelinated dural upward and sealed the edges close to complete the thecal sac and nerve marrow separation. After that, the pediatric surgery and general surgery team separated the rectal, and the plastic surgery team sutured the skin to protect the organ structure.

The third stage was performed when they were nine months old. We closed the sigmoid colostomy. By now, the two sisters were living separately for two months. The overall post-operation condition was good. There were no leakage of spinal fluid and neurologic deterioration.

In order to successfully separating conjoined twins, it relays on all teams understanding how to overcome challenges and difficulties, repeat simulations with the model made from the image, and with the medical and nutritional support and performing bold but cautious phased surgeries. The death rate for emergency separation of conjoined twins is up to 80%. The foreign experts suggest that the conjoined twins will have 70% of survive rate if separated between six to nine months.

Our team learned from the wisdom of related teams in the world and completed the separation of the Rose Sisters when they were seven months old. The keys to successful separation in neurosurgery are: 1) to cut the nerve part prior to intestinal part can avoid infection from the surgery. 2) To cut the nerve system evenly to ensure the best nerve function for the two individuals in the future. We are very thankful for the support from the Master and the foundation, the hospital’s organization, all division’s teamwork, and volunteers’ care. Seeing the Rose Sisters joyfully returned to the home of Tzu Chi members, all our hard works are worth it. We are truly grateful to have this opportunity to utilizing our professional skills and accompany the Rose Sisters to grow.