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Separation of Conjoined Twins : Children's Hospital Experiences

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Nine conjoined (Siamese) twins were separated at the Children's Hospital, Bangkok, Thailand during the period between 1956 and 1995. Six cases were Omphalopagus type of conjoined twins, and the other three were Thoracoomphalopagus type.

Thirteen children survived but five infants expired. Two infants died during the surgical attempt to separate. The other three infants expired after successful separation (thoracoomphalopagus conjoined twins) due to severe cardiac anomalies.

As a result of our own experiences and a review of the literatures, delaying separation till late infancy of these conjoined twins were preferred. In 1989 more advanced investigations were introduced in addition to the routine preoperative studied. The fused visceral organs and the cardiovascular system were evaluated by computerized tomography or 3-dimensional computerized scan and other appropriate hemodynamic measures. These advanced techniques demonstrate the cardiovascular system more accurately which would allow more careful planning of a more complex separation of the twins.

The success of the operation in our series depends on the closed co - operation between pediatricians, nurses, radiologists, the anesthesiologists and the surgeons. Multidisciplinary approach with rehearsal of the events are also very essential.

Conjoined twins is one of the most interesting medical phenomenon. They vary from two symmetrical well-developed individual separated by skin connection to large area of body's region attachments or some internal organs of the body shared by each other.

The world knows the original Siamese twins by the name of Eng and Chang[1] (Figure 1). The Thai prefer to call them Inn and Chand, meaning similar. They were born in 1811 from a Siamese mother and Chinese father who lived in Samut Song-kram province. In spite of their abnormality and of long and difficult labour which preceded their birth, both parents grew genuinely fond of the children. The fact that they were joined together through the lower chest and upper portions of their bellies did not prevent them from developing an active mode of living. When they grew old enough their parents encouraged them to swim in the Maeklong river which was in front of their house. They were found to be useful in helping to look after the ducks and in assisting with household chores. Because of their close anatomical union each was required to concede to the other on many occasions. Eng (Inn) was usually the more aggressive of the two and not infrequently the mother was called upon to make peace between them.

In 1824 Mr. Robert Hunter, a British merchant seeking Thai goods, found his way into Siam. This English gentleman saw at once the great commercial possibilities in the thirteen year-old twins and was not long in setting up a financial agreement with their parents to take the twins abroad. In 1828 an American ship, the Sachem, arrived in Bangkok and the twins, along with their English possessor, set out to see and be seen by the world.

Fig. 1 Original Siamese Twins (Eng-Chang)

They traveled half-way around the globe and were presented at the Royal College of Surgeons, England. The distinguished members and guests of this well known organization examined the first known, living twins of this type with great medical interest. They announced that the twins nervous systems were separated but that they possessed a common naval.

The band connecting the twins had a circumference of 8 inches and was flexible. The British physicians were amazed by the freedom of movement that this union allowed. The children could stand up and lie down back to back. They could run, jump, and ride horseback.

From England the twins toured various European cities. While in Paris rumors spread that impeding mothers should not grance at them for fear children might be affected in the same way!

Because it was the land of free enterprise and unbounded freedom, the twins found their final home in America. There, for some years, they earned their living by appearing on exhibition at P.T. Barnum's museum. This traveling show displayed the twins in many cities and towns across the continent. However, on June 7, 1839, the twins forsook this means of livelihood to settle down in the remote frontier town of Whikesboro, about 60 miles west of Mount Airy, deep in the Blue Ridge foothills. There the "United Siamese Brothers" thrived, raised families, and lived out their lives.

Their family lives are worth mentioning. A Quaker of Dutch and Irish ancestry, named Yates, a farmer by occupation and preacher by action, had two daughters, Sarah and Adelaide. As determined by their horoscopes, Chand (Chang) married Adelaide and Inn (Eng) married Sarah. This double wedding took place in 1843 and was cheered by the parents of the girls and by their friends.

After their marriages the united Siamese brothers moved to farms in White Plains, North Corolina, and they built two houses a mile apart. Perhaps encouraged by the example of strict Quaker fairness, the twins established a region

rule that three days of every week to be spent in one household, three days in the other. This rule was adhered to, winter and summer, through all their lives. The children of Inn (Eng) and Sarah numbered seven boys and five girls. Those of Chand (Chang) and Adelaide were seven boys and three girls.

The Siamese say that one can try to escape anything except one's death. One day in January, 1874, Chand contracted a severe cough accompanied by a fever. As a result of this infection he shortly died. Dr. Joseph Hollingworth of Mount Airy was called in to consider the possibility of separating twins.

At that time modern surgery was still in its infancy. Lister of England, Von Mickulitz of Germany, Kocher of Switzerland, the Mayos of America, were busy in other fields and none had had time to devote themselves to study of surgery of the liver which the twins commonly owned. Dr. Hollingworth decided against any attempt to separate the twins and thus missed the opportunity of being the first known practitioner to attempt separation of Siamese twins.

CHILDREN'S HOSPITAL EXPERIENCES

The first twins (Wandee - Sriwan)

On November 12, 1954 the female conjoined twins were born alive, after eight months of pregnancy by Caesarean section in the Women and Children's Hospital, Bangkok. The development of the twins progressed normally until the age of thirteen months. Then Dr. Sem Pring-Puang-Geo and his surgical staffs decided that an attempt separation of the twins should be made.

Physical examination revealed the twins being united from the tip of the breast bone to the navel, having a conjointed liver but separate abdominal organs, GI tracts, two gallbladders, all of which could be visualized by X-rays. GI contrast studied and IV pyelography.

The operation was preformed on January 20, 1956, through the vertical incision at the skin joining the twins. Further careful incision revealed that the twins shared a common celomic cavity through which the conjoined liver was visualized. A round ligament of the liver with its sleeve was identified and found to run to the point of the conjoined liver. Two distinct gallbladders and GI tract were found. The conjoined liver was then divided vertically. Bleeding from small and large vessels was controlled by fingers pressure and large mattress sutures were made following the entire liver cut surfaces. Satisfactory hemostasis was obtained without much difficulty. After separation of the liver, the fused xiphoid cartilage was cut, pleura of each twins were identified and preserved. Skin and peritoneum composing the opposite wall of the first cut through skin was divided and peritoneal cavity of each twins were packed. The twins were then completely free from each other. No other abnormality of the abdominal organs was found on examination. Both abdominal cavities were closed with undue tension. Post operative response of the divided twins were completely uneventful.

These twins was also the frist successful attempt separating Siamese twins to be performed in this part of the World[1].

The second twins (Prachin - Buri) and the third twins (Suvannee - Sripum)

They were also the omphalopagus type of conjoined twins and were operated at the age of 18 and 17 months with successful separations (Table 1).

The forth twins (Srisakol - Srinakorn)

On June 30, 1963 another living female conjoined twins were born in a village of Sakolnakorn province, North-East of Thailand. They were brought to the Sakolnakorn Provincial Hospital and finally were tansferred to the Children's Hospital. The twins were the children of the third pregnancy. The two former pregnancies resulted in death of the male baby of one month old and a female baby of also one month old.

On admission to the Children's Hospital the combined weight of the twins was 2500 grams, a distinct premature weight.

Because of their "Prematurity" the twins were valnerable to the common gastro - intestinal disease of infancy in this country.

Table 1 Separation of Nine Conjoined Twins in Children's Hospital, Bangkok.

No	Year	Sex	Type	Age at Separation	Shared Organs at Point of Conjoining	Indication of Separation	Result	
							Twin 1	Twin 2
1.	1956	F	OMPH	13 Mon.	- Liver Bridge - Xiphoid Cartilage	Elective	S	S
2.	1961	F	OMPH	18 Mon.	- Liver Bridge - Xiphoid Cartilage	Elective	S	S
3.	1962	F	OMPH	17 Mon.	- Liver Bridge - Xiphoid Cartilage	Elective	S	S
4.	1962	F	OMPH	1 1/2 Mon.	- Liver Bridge - Xiphoid Cartilage	Death of Twin 2.	D	D
5.	1973	F	TH-OMPH	2 yrs.	- Liver Bridge - Common Ventricle - Sternum	Elective	S	D
6.	1989	F	OMPH	3 1/2 Mon.	- Liver Bridge - Lower Part of Pericardium - Xiphoid Cartilage	Elective	S	S
7.	1991	M	TH-OMPH	8 Days	- Liver Bridge - Pericardium - Sternum - Xiphoid Cartilage	Deterioration of Twin 2	S	D
8.	1993	M	TH-OMPH	2 Days	- Liver Bridge - Pericardium - Sternum - Xiphoid Cartilage	Deterioration of Twin 2	S	D
9.	1995	M	OMPH	5 Mon.	- Liver Bridge - Xiphoid Cartilage	Elective	S	S

In spite of all doctor's and nurse's attempts to save their lives, condition of the twins deteriorated. Diarrhea and restlessness worsen and finally one of the twins (Srinakorn) succumbed at 0.35 A.M. of August 13, 1963.

Surgical attempt to separate the living one (Srisakol) from the dead one was started at 1.45 A.M. on August 13. 1963. At the time of the death of one of the twins (Srinakorn) the other one (Srisakol) was still in good condition and was able to withstand surgery undertaken one hour and 10 minutes later.

In order to save the life of the living one, skin incision of the bridging portion was taken over a little bit more on the non-living side.

On entering into the peritoneal cavity there was foamy fluid on the non-living side while the peritoneal cavity of the living one looked glittering and healthy. The only organ that joined together was the liver which was divided in the conventional technique. It took 45 minutes to complete the separation.

After operation the short-lived Srisakol expired after having respiratory failure one hour later.

Because of the immediate onset of anemic looking appearance of Srisakol and because of the result of operative finding of good bleeding

when the bridging liver was severed and autopsy findings, we assumed that the cause of death of Srisakol 3 hours after the death of her sister was the loss of blood through the bridging liver into the dead side. While living, blood pressure in each one of the twins, eventhough not really equal, but being well pressuring so that blood of one could not flow into the other.

The fifth twins (Kate - Kao)

The twins were born on December 15, 1980 in Prachinburi province, central of Thailand. They was transferred to the Children's Hospital at the age of two days. The examination revealed thoraco-omphalopa-gus type of conjoined twins. X-rays, including contrast studies revealed separate GI tract, GU tract and a common shadow of liver shared between them. Two distinct gallbladders were visualized. Cardiac catheterization showed the common connection between left ventricle of both twins. The development of these twins progressed normally until the age of 2 years, which was the appropriate time for separation.

On opening into the peritoneal cavity, the bridging liver was divided in the same techniques as being performed in the twins No. 1. The operative procedure was extended into the thoracic cavity and the channel between left ventricle of both twins was divided and closed. Both abdominal cavities were closed without any difficulty. Because of the absent sternum, the chest wall of both sides of the twins were then closed with the use of prosthetic sheets.

Postoperative response of Kate was uneventful, but Keo expired after having difficult breathing and postoperative myocardial dysfunction two hours later. Autopsy findings revealed common atrium and ventricle, pulmonary atresia and patent ductus arteriosus.

The sixth twins (Vullee - Viravan)

The female living omphalopagus type of conjoined twins were born in Suraj-Thanee Provincial Hospital, southern of Thailand, on January 5, 1989. They were brought to the Children's Hospital at the age of 13 days (Figure 2).

Abdominal ultrasonography confirmed two kidneys in each infant with no evidence of hydronephrosis. Computerized tomography scan confirmed a fused liver occupying in both peritoneal cavities (Figure 3).

The hearts and vascular systems were stud-

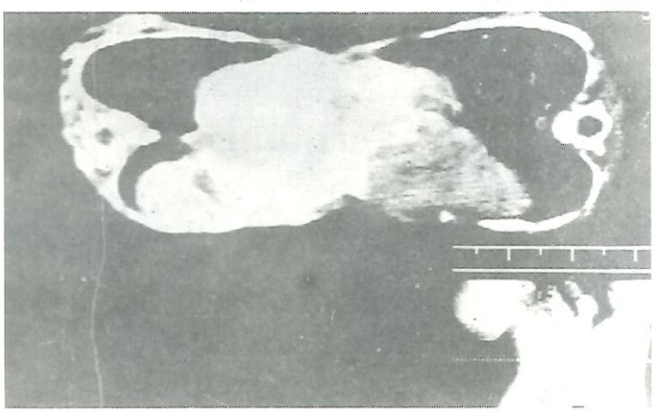

Fig. 3 CT scan of abdomen in Twins No. 6.

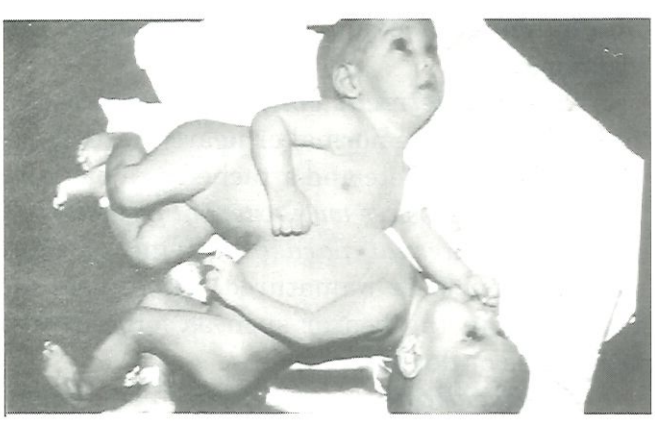

Fig. 2 Twins No. 6 (Vullee - Virawan).

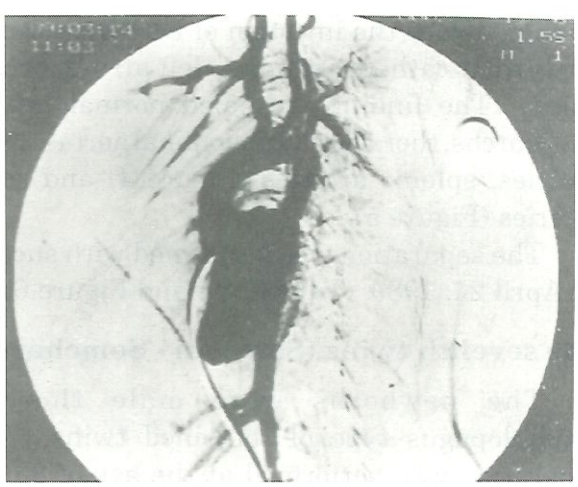

Fig. 4 Heart, aortic arch, thoracic aorta, celiac and splenic arteries of each child of Twins No. 6.

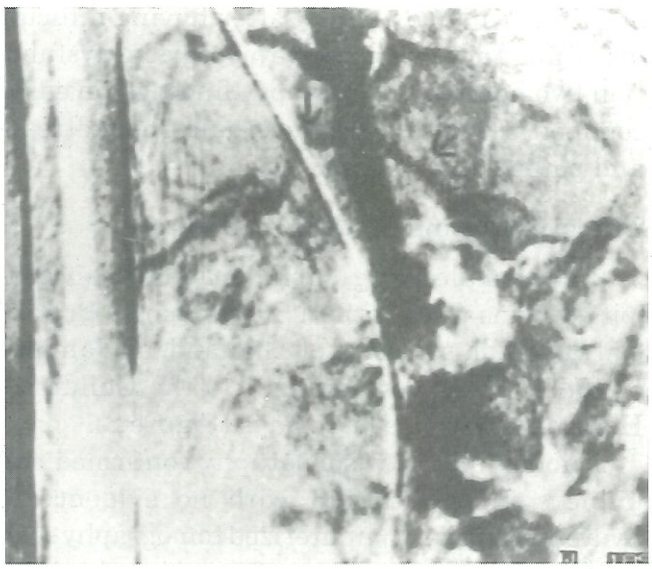

Fig. 5 Aortogram showing normal renal arteries in each of Twins No. 6.

Fig. 6 Twins No. 6 after separation at age of 6 months.

ied by using 3 - dimensional computerized scan combined with the injection of DISIDA through the cardiac catheterization in left atrium of both twins. The findings revealed normal hearts, aortic archs, thoracic and abdominal aortas, celiac arteries, splenic arteries (Figure 4) and renal arteries (Figure 5).

The separation was performed with success on April 24, 1989. (Tables 1, 2 and Figure 6).

The seventh twins (Somchai - Somchart)

The newborns were male thoraco-omphalopagus type of conjoined twins. The separation was performed at the age of 8 days because of the deterioration from heart failure of one twins (Somchart). Postoperative recovery of one twins (Somchai) was uneventful, but Somchart expired one hour later. Autopsy findings revealed coarctation of aorta, transposition of great vessels, VSD, ASD and PDA.

The eighth twins (Top - Ten)

The eighth twins were male thoraco-omphalopagus type of conjoined twins. The separation was attempted at the age of 2 days with the same indication as the former twins. *Top* survived but *Ten* died two hour later. The report on autopsy of Ten revealed ASD, PDA and pulmonary hypertension (Tables 1,2).

The ninth twins (Chaiwat - Chatchaval)

They were also of omphalopagus type of conjoined twins. Only conjoined livers and xiphoid process were detected on the series of investigations. The operative separation was performed at the age of 8 months. Both children survived and well.

DISCUSSION

Conjoined twins occur approximately once in 50,000 births,[1-3] but the incidence in Thailand seems to be more often than in the other part of the world which is 1:34,000.[1] The majority presented with thoracoomphalopagus union 73 per cent, pygopagus (conjoining at sacrum and coccyx) 19 per cent, ischiopagus 6 per cent and craniopagus 2 per cent.[2]

Conjoined twins are classified into three groups according to the parts of the bodies which are joined and shared.[2-5]

1. Symmetrical conjoined twins. Each component of the twins is complete or nearly complete. These groups are thoracopagus, omphalopagus, pygopagus, ischopagus and craniopagus.

2. Asymmetrical conjoined twins. One individual may be normal or nearly normal and the other incomplete and attached as a parasite.

2.1 Autosites with external parasites.

2.2 Internal parasites. One or more parasitic twins or immatured cell tumors are found within the bearer such as fetus-in-fetu, teratomas or dermoids.

3. Miscellaneous. These groups are polydactyly or gastrointestinal duplication.

Etiology of the conjoined twins is still not known. Various theories of embryologic deviation and mechanism have been suggested by many authorities. Anatomist and embryologist believe that identifical twins as well as conjoined twins arise from a single zygote. Incomplete duplication or separation of zygote during the period of blastocyst (20th postovulatory day) result of the various types of conjoined twins.[4,6]

According to data compiled from the literatures about the sharing organs of thoracoomphalopagus, liver fusion is found in 80 per cent,[7] attachment of pericardial sac in thoracopagus 90 per cent and 25 per cent of these have conjoined heart.[8]

Almost all of the cases are diagnosed and treated after birth. Occasionally the diagnosis was made before delivery. Caesarean section is therefore the procedure of choice to assure maximal care in delivery (Twins No. 1). Plain film and ultrasonography presented the following criteria for prenatal diagnosis of conjoined twins (1) boths may be in breech positions, (2) usually there is hyperextension of cervical spines, (3) 50 per cent associated with polyhydramnios[2] which can be detected as early as 12 weeks of gestation[9].

Many reports indicated a preference for delaying operation till late infancy[2,4,10] (Table 1). Kiesewetter[11] reported the separation of thoracopagus twins at 12 days age, who were joined at pericardial septum and liver. One child survived and the other one died. A similar case was reported by de Vries[12] as successfully separated at the age of 6 months. However, for the life-threatening emergencies that must be treated by immediate separation of the twins are ruptured omphalococle,[13] intestinal obstruction,[14] jejunal atresia[15] and early deterioration of one or both twins[16] (Twins No 4,7,8).

The extent of the preoperative studies to be carried out before attempts at separation of conjoined twins varies from case to case beyond rountine preoperative assesment. The fused visceral organs and cardiovascular system should have been evaluated by computerized tomography and 3-dimension computerized scan[15] which can demonstrate the anatomical relationship of cardiovascular system more accurately (Twin No. 6,9).

When the union includes only the superficial structures such as skin, subcutaneous tissue and occasionally only the cartilage, successful separation is very high. Koenign in 1689 performed the first successful separation of a skin tube joining Siamese twins at the area of navel by simple ligation[17]. In thoracopagus twins in which 25 per cent have conjoined heart[8], anomalies in the cardiopulmonary system may be so extensive as to make separation impossible or only one individual may survive (Twins No 5, 7, and 8). Separation may also be complicated by absent or shared of biliary systems.[18]

Experiences of medical teams (pediatricians, nurses, anesthesiologists, radiologists and surgeons) and the life-supporting equipments are of utmost importance. Mulcare[19] in 1970 separated a 10 months old xiphopagus (omphalopagus) conjoined twins at Nakhon-Ratchasima Provincial Hospital in North - eastern Thailand resulted in only one survival. The other one died of respiratory complication and the lack of adequate respiratory suppost equipment.

Anesthesiologists also play a significant role in the success of surgical separation. According to prolong operation, all physiologic measurement in both babies must be made to ensure adequate maintenance throughout the operation. Any blood loss must be carefully calculated and transfused to both twins.[20]

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