

Successful Surgical Separation of Craniopagus Twins - A Monumental Achievement in the History of Bangladesh

MAA SALEK^a, M HASAN^b, MAHMED^c, AA BEAUTY^d, MA ISLAM^e, MA KALAM^f

Abstract:

Craniopagus is a condition of conjoined twins that are fused at the cranium. After birth, 25% of craniopagus twins survive and can be considered for surgical separation. It is a complex multistage, multidisciplinary team involvement exercise needing detailed evaluation, planning and preparation beforehand. A harmonious collaboration from the clinical, administrative as well as logistic point of view is mandatory for successful management of such complex condition.

The honorable prime minister of Bangladesh picked up “Rabeya and Rukaiya”, a craniopagus twins, the first ever case in the history of Bangladesh and handed them over to

an expert team from Hungary and Bangladesh for successful management. The noble joint exercise known as “Operation Freedom” took place in home and abroad through multiple procedures/interventions in different institutions by a group of experienced panels of medical and non-medical professionals. In this article we will highlight the events of this monumental achievement.

Keywords: Craniopagus, conjoined twins, multidisciplinary team, operation freedom, expert panel

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Introduction:

Craniopagus twins (CPT) are a rare congenital malformation or dysmorphism in which twins are conjoined and fused at the cranium, accounting for only 2%–6% of conjoined twins, with an incidence once in every 0.6 to 2.5 million lives births.¹ CPT is different from craniopagus parasiticus in which a parasitic twin head with an undeveloped body is attached to the head of a developed twin.

Females are more predisposed than males with a ratio of 3:1 although this has not been correlated with any cause

of the condition.² Mortality rate of CPT is 40% stillborn and 33% die in perinatal period.³

The skulls are often joined in homogeneous regions on each twin in both vertical and angular directions.⁴ Conjoined brain tissue, connected arteries and veins, and defects in the skull and Dura make surgery technically challenging and difficult.⁵ Although there is high prenatal and postnatal mortality for CPT, successful separation has become more common due to advances and improvements in neuroimaging, neuro-anesthesia, and neurosurgical techniques.⁶⁻⁷

Neuroimaging, including CT, MRI, and conventional angiography, plays an important role in mapping the connected arterial and venous structures, brain parenchyma, calvaria, and Dura.⁸⁻⁹

Understanding the shared vascular anatomy is important for surgical planning because separating common vessels is associated with complications and manifestations such as thrombosis, pulmonary embolism, infarction, and hemorrhage.¹⁰

Separation can take place in single or multistage procedures and has improved as the understanding of the physiology, surgical techniques, and technology of CPT have evolved.¹¹

Moreover, Separation of conjoined twins poses several technical, legal, religious, moral and ethical issues.¹²

- a. Md. Al Amin Salek, Department of Neurosurgery, Combined Military Hospital Dhaka,
- b. Masroor Hasan, Department of Plastic Surgery, Combined Military Hospital Dhaka,
- c. Masud Ahmed, Department of Neuro-anesthesiology, Combined Military hospital Dhaka,
- d. Anjuman Ara Beauty, Department of Pediatric Neurology, Combined Military Hospital Dhaka,
- e. Md. Shafiqul Islam, Department of Neurosurgery, Dhaka Medical College Hospital,
- f. Md. Abul Kalam, Director, Sheikh Hasina National Institute of Burn and Plastic Surgery

Address of Correspondence: Md Al Amin Salek, Associate Professor and Classified Neurosurgeon, Department of Neurosurgery, Combined Military Hospital, Dhaka Cantonment, Bangladesh. Email: salek1972@yahoo.com

In this write up we will highlight the events of “Operation freedom” as a monumental achievement in the history of Bangladesh.

Case report

Conjoined twin, Rabeya and Rokeya was born in Pabna in 2017 to a couple who are schoolteachers. They got admitted into Combined Military Hospital Dhaka on 22 July 2019 for final separation of craniopagus (Fig-1).



Fig-1: “Rabeya and Rukaiya” as craniopagus twins with their parents

Both the babies who born with fusion in the head region were healthy after birth. Their developmental milestones were age appropriate. After a detailed clinical and radiologic assessment, decision was made to perform the separation by planning a multistage, endovascular and operative interventions with multidisciplinary team.

They underwent multiple interventions in two sessions at home and abroad by groups of specialized teams before getting admission to our hospital.

In initial session they underwent endovascular interventions for vascular separation of common pseudo- sinus of the brain (novel technique in the craniopagus management) at Dhaka Medical College Hospital, Bangladesh.

Later as a second session they underwent multiple plastic surgery procedures with insertion of subcutaneous tissue expanders which were aimed to expand the skin of the scalp so that it can be comfortably closed after the separation in Hungary.

Ultimately the final session of surgical separation of brain and the coverings was done in CMH Dhaka.

Operation Freedom- Events

The final stage of surgical separation of these craniopagus twins was named “operation freedom”. A specialized group of multidisciplinary teams from home and abroad could work relentlessly and comfortably by

the mammoth logistic arrangements and extensive administrative support under the leadership of the commandant hospital.

Operation freedom started at 0400 hours on 01 August 2019 and it took a long 33 hours by a multidisciplinary team of anesthesiologists, neurosurgeons and plastic surgeons from Bangladesh-Hungarian origin.

1. Support

The honorable prime Minister Sheikh Hasina and her dedicated team showed maximum empathy for them and kept open all avenues for their best possible management. (Fig-1)

2. Medico legal issues:

After a detailed clinical and radiologic assessment, we decided to perform the separation by planning a multistage, endovascular and operative project with our multidisciplinary team with proper medicolegal, religious and bioethical considerations.

3. Coordination:

During the entire process, the concerned doctors and cooperating international participants were centrally coordinated to achieve maximal sharing of information and quick and proper logistics.

4. Sessions:

The first session of the endovascular phase was performed in Dhaka, Bangladesh, in February 2018.

The plastic surgery phase aimed to implant silicone; subcutaneous tissue expanders was performed in Hungary for a period of seven months.

The final session of separation surgery took place in CMH Dhaka on August 1 and 2, 2019. The separation procedure took 33 hours; 4 neurosurgery, 4 plastic surgery, and 4 anesthesia teams worked in rotation during the operation. (Fig-2)



Fig-2: Activities in Operation Theatre. Operation in progress

5. Post operative Management:

After regaining consciousness and reaching a stable general condition, one of the twins was weaned from the ventilator and then extubated on the sixth postoperative day. The weaning of the other child was only possible in the sixth week. They were managed by a multidisciplinary team from home and abroad.

6. Post of complications

Rukaiya had a hectic post op period with unstable haemodynamics, severe sepsis, fluctuation of biochemical parameters. She had developed a massive spontaneous intracranial hemorrhage (ICH) for which she underwent emergency exploration and evacuation of ICH.

7. Post operative Neuro-rehabilitaion

Comprehensive neuro-rehab care including LASER, Helmet, Physiotherapy, Speech therapy and Occupational therapy by an expert team of home and abroad.

8. Final Briefing

A friendly international press briefing was held to disclose the events, outcomes, and future plan.

9. Follow up

Rabeya is activities of daily life (ADL) independent. Rokeya is ADL dependent. She can sit and stand with support; has a lessening degree of spasticity in her extremities; can do purposeful arm, hand, and head movements in an inconsistently reproducible manner; and can be fed orally (Fig-3)



Fig-3: *the children after operation in a good health*

Discussion:

Craniopagus twins occur in 10 to 20 cases per billion births. These twins are of the same gender and a female preponderance has been noted (3:2).¹³ The first partially successful separation of craniopagus twins was carried out in 1952 by a team of neurological surgeons led by Dr. Oscar Sugar. Even though both the twins survived the operation that lasted for twelve hours, only one regained consciousness postoperatively while other remained in a coma for 34 days and eventually expired.¹⁴ On the basis of review of 64 cases, Stone and Goodrich¹⁵ defined partial CPT as lacking substantial shared dural venous sinuses, whereas total CPT share a large portion of their dural venous sinuses and present with pronounced brain compression, which leads to distortion within the cranium.

Separation of craniopagus twins is usually an elective, multidisciplinary and staged surgery that involves the well co-ordinated efforts from a multi-disciplinary team. The credit of separation of the youngest craniopagus twins till date goes to Nejat et al., who separated craniopagus twins aged 32 weeks of gestation and weighing 1250 grams each, after the death of one of the twins.¹⁶ Over the last 22 years (1995 to 2017), 20 cases of craniopagus separation have been reported with mixed results.¹⁷ Normally, a partial craniopagus is associated with a better chance of separation. However, Harvey et al., suggested that only the presence of a vertical craniopagus was a statistically significant favorable factor in the successful separation of craniopagus twins.

Table i shows details of the craniopagus twins in chronological order in whom surgical separation has been attempted.

This surgery has generated few salient features. Preoperative evaluation is essential to start the journey of such complex procedure. A harmonious understanding between group of specialist doctors and administrative-logistic support team are mandatory to make it successful.

In case a common and major intracranial dural venous sinus is shared between the craniopagus twins, the possibility of a venous bypass graft must be considered at an earlier stage, prior to the time when the actual procedure to separate the two children is being undertaken.¹⁷ But in our case endovascular embolization

Table-I

Reported data related to the separation of craniopagus twins (1995-2017)

Name	Sex	Age at separation	Type	Surgeon-in-Chief	Year of separation	Number of children alive after 1 month of surgery (the perioperative period)	Single or staged surgery (number of stages)
Sudan twins	M	32 weeks	PA	Nejat F	Unknown	1*	Yes
Hira and Nida Jamol	F	2.25 years	TV	Hoffman	1995	2	Yes
Bessy and Dores Gonzalez	F	11 months	TV	Walker	1996	2	No (5)
Joseph and Luka Banda	M	11 months	TV	Carson	1997	2	Yes
Taylah and Monique Armstrong	F	6 months	TV	Campbell	2000	2	Yes
Ganga and Jamuna Shrestha	F	11 months	TV	Goh	2001	2	Yes
Alyssa and Bethany Nolan	F	18 days	TA	Campbell	2001	1*	Yes
Ghuangzhou twins	F	1.42 years	TA	Unknown	2001	1	Yes
Josie and Teresa Hull	F	1 year	TV	Kawamoto/Lazareff	2002	2	Yes
Laleh and Ladan Bijani	F	29 years	TA	Goh/Carson	2003	0	Yes
Ahmed and Mohamed Ibrahim	M	2.33 years	TV	Salzer	2003	2	Yes
Carl and Clarence Aguirre	M	2.5 years	TV	Goodrich/Staffenberg	2004	2	No (4)
Greek twins	F	4 months	TV	Di Rocco	2004	2	Yes
Florianopolis twins	M	1.25 years	TV	Unknown	2006	1	Yes
Poland twins	M	2 days	TV	Unknown	2010	1*	Yes

of common vascular sharing as a noble procedure could make the surgery less troublesome. To the best of our knowledge, this is the third case of successful incorporation of an endovascular occlusion of the shared venous channels in craniopagus twins, and this is the first successful attempt of a full-segment, endovascular occlusion of a largely shared intracranial venous system in craniopagus twins.¹⁸

A meticulous attention to the patients' positioning during surgery should be given, taking into account that none of the twins is getting compromised as a result of the weight of the other twin or the torsion induced at the site of the craniopagus by an unnatural position.¹⁹ we used two neurosurgery table with special support system to make patients position optimum. It is imperative to plan for an adequate scalp cover for both the children even if it entails using skin expanders, planning an myocutaneous skin flap or a transposed scalp flap using microvascular techniques.²⁰ In our case several sessions of plastic surgery procedures were done to facilitate scalp closure after surgery.

Conclusion:

Successful separation of viable conjoined twins has been historically a great rarity. Successful separation of twins where both have come out of surgery without any neurological deficit remains a rare occurrence and, in our mind, the ultimate goal.

Management of such rare condition needs a multidisciplinary team approach. The application of latest technique and technology together with synchronized skillful approach are considered as prime factors to deal such complex operations successfully. And lastly, we can consider the success of "Operation Freedom" as a monumental achievement in the history of medical science of Bangladesh.

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