

# Gemini Untwined: treating craniopagus conjoined twins

BY SOHAIB R RUFAT, SRI GORE AND NOOR UL OWASE JEELANI

The authors discuss the successful separation of craniopagus conjoined twins at Great Ormond Street Hospital and the role of the ophthalmologist in such cases.

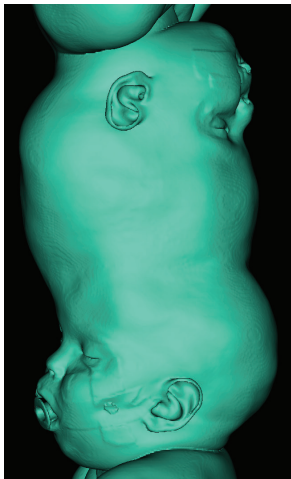


Figure 1: Three-dimensional soft tissue reconstruction demonstrating craniopagus malformation.

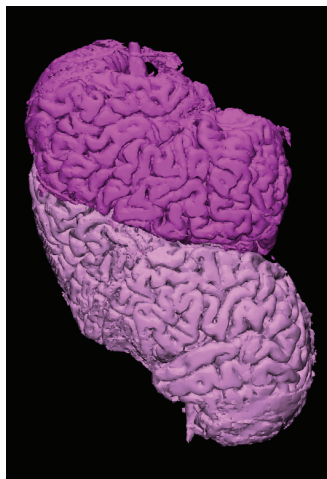


Figure 2: Three-dimensional reconstruction of the brains of the craniopagus twins.



Figure 3: Tissue expanders in situ to produce sufficient skin for separation and craniofacial reconstruction.

Craniopagus conjoined twins are extraordinarily rare, occurring in only one in 2.5 million births and representing only 2-6% of conjoined twins [1,2]. Rarer still are male craniopagus twins, representing only 30% of craniopagus twins [2]. Sadly, around 40% of craniopagus twins are stillborn or die during labour, while a further third die within 24 hours [2]. These highly complex cases present unique challenges to the entire multidisciplinary team, including the ophthalmologist. As these cases are so rarely encountered, it is helpful to share experiences and lessons learned for each case.

## Yigit and Derman

On 21 June 2018, conjoined twins Yigit and Derman were born in Antalya, Turkey [3]. They were delivered by caesarean section at 32 weeks. Their father had built a special crib which could accommodate the twins, who were unable to face the same direction or face each other [3]. Their development and quality of life was severely affected by their condition, but surgically separating such twins represents an extremely difficult task, often associated with devastating results. Harvey and colleagues [4] reviewed

14 attempted surgical separations from 1995 to 2015 and found that over a third of cases ( $n=5$ ) were unsuccessful, either resulting in the death of both twins ( $n=1$ ) or one twin ( $n=4$ ). Multi-stage operations were associated with the highest chance of success [4].

On 2 December 2019, the twins were referred to Great Ormond Street Hospital (GOSH), London. The treatment at GOSH was funded by Gemini Untwined charity [5]. They were diagnosed with total vertical (Type 2) craniopagus malformation, meaning that the two twins were joined at the top of the head and faced approximately 180 degrees away from each other, as shown in Figure 1. The two brains were merged together, as can be appreciated in Figure 2, with multiple shared venous sinuses and arterial crossover vessels. It was decided that the safest way to proceed was with staged surgical separation, featuring the insertion of tissue expanders to produce enough skin to cover the two scalps, followed by three procedures to separate the brains and blood vessels and reconstruct the skulls.

## Modified ophthalmic examination

The ophthalmic examination was challenging, as conventional methods were

not feasible for this case. Our modified approach is described in detail in our recently published paper in the *Journal of Surgical Case Reports* [6]. Specifically, we adopted a modified approach using forced-choice preferential looking, portable slit-lamp biomicroscopy, indirect ophthalmoscopy and optical coherence tomography (OCT).

To the best of our knowledge, this represents the first use of handheld OCT in craniopagus twins [6]. We used the Envisu C2300 handheld OCT device (Leica Microsystems, Wetzlar, Germany) to examine the optic nerve heads and foveae (Figure 4). Handheld OCT findings were normal in both twins [6].

The main ophthalmic abnormality was due to high right brow positions in both twins secondary to scalp shortage. This caused right exposure keratopathy secondary to lagophthalmos in both twins, which could be appreciated well during examination under anaesthesia using the Keeler Portable Slit-lamp (Keeler Ltd, Windsor, UK). This was successfully treated with topical lubricants but will likely require full correction with surgery in the future.



Figure 4: Handheld OCT image acquisition in operating theatre.



Figure 5: Twins post-separation surgery, pictured undergoing physiotherapy.

## Surgical separation

The twins were separated on 28 January 2020 (Figure 5) and discharged from GOSH on 9 June 2020 back to Turkey. This case was covered in a Channel 4 documentary, *Conjoined Twins* [7]. This represents the fourth separation of craniopagus twins at GOSH and the largest global experience of successful separations of craniopagus twins.

The surgical treatment was supported by detailed neuroimaging, 3D modelling and virtual reality simulation. Over 100 professionals across more than 15 disciplines contributed to Yigit and Derman's treatment. After five weeks of tissue expansion, their three surgical separation procedures totalled 36 hours of surgery and the entire surgical process took seven weeks – quicker than the previous three sets separated at GOSH. This minimised the risk of complications due to a potentially prolonged hospital stay. Following separation, right brow positions grossly improved in both twins.

## Reflections

It is extremely rare for an ophthalmologist to be asked to assess craniopagus conjoined twins – indeed, most ophthalmologists may never see such a case throughout their careers. Notably, we have learned that handheld OCT is feasible and clinically valuable in such cases, not only to provide a detailed baseline assessment of the optic nerve heads and foveae, but also to monitor and exclude any potential pathology during the course of treatment. Indeed, when one twin displayed altered visual behaviour in the postoperative period, the handheld OCT excluded any new retinal or optic nerve pathology. Furthermore, assessing vision in young preverbal children is challenging, so the ability to examine structural morphology using serial handheld OCT examinations is clinically valuable.

As luck would have it, in August 2019, Noor ul Owase Jeelani, Senior Neurosurgeon, was holidaying with his family in Antalya, Turkey – the hometown

of Yigit and Derman, where he was able to review them and speak with the family and the Turkish medical team. The case had been referred to him four weeks previously [5]. He and his colleague, Professor David Dunaway, Professor of Craniofacial Surgery at GOSH, had just started their charity, Gemini Untwined, after recently separating Pakistani craniopagus twins, Safa and Marwa [8]. Gemini Untwined was able to raise the necessary funds and mobilise resources to help Yigit and Derman [5].

The main focus of Gemini Untwined is to treat craniopagus conjoined twins, but the associated research, technology and expertise can be applied to treating more common craniofacial abnormalities. Gemini Untwined has brought together leading clinicians, scientists, engineers and other professionals with one common goal – to defy the odds and give craniopagus twins the chance to return home as two separate, healthy children.

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