Craniopagus: Overview and the implications of sharing a brain

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Abstract

Craniopagus twins, who are conjoined at the head, are uncommon and often misunderstood. While craniopagus is rare in itself, Krista and Tatiana Hogan are unique even among craniopagus twins: their brains are connected. In this review, I will explore the history of craniopagus as well as our current understanding of the malformation. Furthermore, I will discuss surgical separation techniques, classification systems, and how these have led to higher survival rates in separated craniopagus twins. Surgical separation of craniopagus twins is perhaps the most formidable of all neurosurgery operations, particularly in the presence of shared neural tissue. Krista and Tatiana fall into this daunting category. The risk of neural damage, coupled with circulatory complications, led Krista and Tatiana's physicians to conclude a separation would be too dangerous. Consequently, Krista and Tatiana are left with a connection that is both novel to documented research and exquisitely mysterious. They possess what their pediatric neurosurgeon Dr. Doug Cochrane has called a "thalamic bridge" (Dominus, 2011a). Krista and Tatiana's thalamic bridge will provide significant insight into the study of cognition and behaviour, and may even have significant implications to the philosophy of mind. Furthermore, their connection will be accompanied by major social change, as we must redefine our definition of what it means to be an individual. For us to understand how being part of a pair is more important than being an individual to one's identity, we must shift our perspective and eliminate our preconceived notions of individuality.

Keywords: *craniopagus, conjoined twins, thalamus, brain, surgical separation*

Examples of human fascination with twins are evident through history. The Gemini constellation, known in Greek mythology as Castor and Pollux, is arguably the most well known set of twins. Castor and Pollux fought great battles alongside Herkales and others in what became known as the Argonauts (Hard, 2004). The Greeks held these twins in high standing: they were not only great warriors, but raised by gods. Conjoined twins, though less recognized, also appear in Greek and Roman mythology. Kteotos and Eurytos, known as the Molionides, were joined at the hip and adversaries of Herkales. The Molionides destroyed Herkales's armies and are responsible for

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one of the great warrior's rare losses in battle (Hard, 2004). Furthermore, Janus, the Roman god of beginnings and transitions, is depicted as having two faces. Initially Janus may not resemble conjoined twins; however, a recent interview with nineteen-year-old brothers Stefan and Tyler Delp, craniopagus twins facing opposite directions, shows just how similar Janus is to conjoined twins (Hochman, 2011). Greek and Roman mythology clearly glorify twins, and to this day, twins continue to captivate us.

Krista and Tatiana Hogan are craniopagus conjoined twins. Craniopagus twins, a term originally coined by August Förster to describe twins conjoined at the head (as cited in Browd et al., 2008), are a rare (i.e., 0.6 per million births; Bucholz, Yoon, & Shively, 1987) and misunderstood congenital disorder, representing only 6% of all conjoined twins. If born, the chances of craniopagus twins surviving past infancy are quite low (Stone & Goodrich, 2006): 40% are still-born (Browd, Goodrich, & Walker, 2008) and 33% die within twenty-four hours of birth. Despite these harrowing odds, Krista and Tatiana Hogan are already beyond their fourth birthday. They not only survived, but they are healthy and thriving.

While craniopagus is rare in itself, Krista and Tatiana are unique even among craniopagus twins: their brains are connected. Krista and Tatiana's neural bridge will provide significant insight into the study of cognition and behaviour, and might even have significant implications to the philosophy of mind. Their unique neural connection may forever change the meaning of individuality and perception. In this review, I will explore the history of craniopagus as well as our current understanding of the malformation. Through this we can begin to grasp what Krista and Tatiana's lives will entail. From them, we must gain an appreciation of how their brains are connected, and objectively contemplate what it means to be an individual.

Modern History of Craniopagus

Although there are cases of conjoined twins dating to the 10th century (as cited in Browd et al., 2008), it was not until 1491 that the first case of craniopagus was documented (Browd et al., 2008). Sebastian Münster's famous work *Cosmographia universalis* provides the first account of craniopagus twins (as cited in Walker & Browd, 2004). While discussing Bavarian history, Münster mentions a pair of unique twins, joined at the head, who lived for ten years. Münster follows with an account attributing the twins' malformation to a punishment for their mother's mistakes (as cited in Browd et al., 2008). Furthermore, Browd et al. (2008) discuss a chapter from "On Monsters and Marvels," a republished book, originally written by 16th Century French surgeon Ambroise Paré. In the chapter entitled "Monsters," Paré depicts various types of "supernatural" twinning, some of which, curiously enough, resemble craniopagus. Later, Paré asserts that "[i]t is certain that most often these monstrous and marvelous creatures proceed from the judgment of God…" (p 5). Fortunately, over time, this radical view towards conjoined twins began to change.

Chang and Eng, the original "Siamese twins," embraced their malformation. They traveled in road shows near the beginning of the 19th Century – eventually becoming not only rich, but a new type of art (as cited in Browd et al., 2008). Nevertheless, because of the rarity of craniopagus twins, even now many still share Paré's view of them as tragic or punished. The Delp family's fear of people's ignorance and discrimination led

them to hide Stefen and Tyler from much of the world. It was not until June of 2011 that they gave their first interview – at the age of nineteen (Hochman, 2011). Stefen, Tyler, Krista, Tatiana, and all other craniopagus twins are so rare that the most recent review shows a total of only 64 documented cases (Stone & Goodrich, 2006). The discussed history of craniopagus extensively shows how craniopagus twins can be seen by some as "monsters." Therefore, combating ignorance through both knowledge and being mindful of existing prejudices are vital steps in removing ancient stereotypes associated with craniopagus twins, thus providing Krista and Tatiana with the social acceptance they deserve.

Classification Systems and Surgical Interventions

In addition to increasing social awareness, there must be a push to increase scientific understanding of separation techniques; and, if separation is not an option, how to improve quality of life for the twins. Over the past fifty years, craniopagus research has had two main focuses: surgical separation techniques and classification systems (Browd et al., 2008; Bucholz et al., 1987; O'Connell, 1976; Stone & Goodrich, 2006; Walker & Browd, 2004; Winston, 1976). Increasing our knowledge of surgical separation techniques and classification systems is of utmost importance to advance research on craniopagus twins; however, addressing the ongoing debate of classification systems should precede any consideration of separation techniques. A secondary research question, which is related to both areas of focus, is the question of how craniopagus twins form. While this is unknown, Spencer (2000a) argues that conjoined twins result from fusion of two embryonic disks. Specifically, Spencer (2000b) states that craniopagus twins may result from fusion of primitive neural folds. Nevertheless, without a concrete cause, prevention is not an option and the focus must be on treatment.

The precision of existent classification systems and their correct application are critical for those attempting to successfully separate craniopagus twins, since different junction sites present different complications (Browd et al., 2008). The first attempt at a craniopagus classification system was made by O'Connell (1976), who refuted the philosophy of classifying according to the site of cephalic junction. Instead, he stated that depth of junction was the most important aspect in determining survival after separation. O'Connell (1976) proposed a differentiation between partial and total craniopagus. Partial craniopagus, characterized by a small cephalic junction, has the possibility that each cranium is still intact. Conversely, total craniopagus would be characterized by a wide connection: two brains, one cranium. Additionally, O'Connell (1976) discusses three different subclassifications of total parietal craniopagus, or as he calls it, vertical craniopagus. Type I entails the twins facing the same direction; Type II entails the twins facing the opposite direction (inter-twin angle ≥ 140°); and Type III entails anything in between.

A decade later Bucholz et al. (1987) contested O'Connell's (1976) system, stating that the difference between partial and total craniopagus described was not precise. Instead, four different classifications were introduced: frontal, parietal, temporoparietal, and occipital. These classifications, unlike O'Connell (1976)'s system, are dependent on the specific location of the junction. Bucholz et al.'s

(1987) classifications are junctions of either the frontal bones, parietal bones, both the parietal and temporal bones, and occipital bones, respectively. Yet, literature on recent separation attempts does not support Bucholz et al.'s view; rather, it reinforces classification according to O'Connell's (1976) system by building on it (Browd et al., 2008; Parameswari, Vakamudi, Raghupathy, & Siddhartha, 2010). In addition to this new classification, Bucholz et al. (1987) also proposed a venous scale to classify the amount of venous drainage interrupted during surgery: (1) no venous drainage was interrupted; (2) only cortical veins were divided; (3) major dural sinuses were encountered and ligated in the course of separation (Bucholz et al., 1987). Venous drainage describes the extent of which a twin's blood may be drained from their brain, collected in a sinus, or reservoir, and then sent to the heart to be re-oxygenated. If this drainage is interrupted, blood may be prevented from leaving the brain, resulting in swelling which ultimately leads to neural damage. This venous drainage classification system, important to subclassify different cases of craniopagus, has the strongest correlation with survival and outcome of the separated twins. Classification of these systems has proven essential in separation attempts, as the presence of a shared venous system is now known to be the single largest risk factor for fatalities (Browd et al., 2008).

Recently, Stone and Goodrich (2006) added to O'Connell's (1976) system by further subclassifying total and partial craniopagus into vertical and angular. More specifically, they define total craniopagus as the twins sharing a significant venous sinus and partial as an absence of a shared venous sinus. They define angular as "an inter-twin longitudinal angle below 140°, regardless of axial rotation" (p 1084). Conversely, Stone & Goodrich (2006) define vertical as having a continuous cranium. Thus, their system is comprised of four major classifications: total vertical, total angular, partial vertical, and partial angular.

The most recent proposed classification scheme comes from Browd et al. (2008), who stress the importance of including important risk factors such as "connectivity between scalp, calvaria, dura mater, neural tissue, arterial and venous connections, deep venous drainage, and ventricular anatomy, as well as atrial and venous outflow" (p 5). They provide a score, between 10 and 28, with a higher value suggesting a more difficult separation.

Classification systems, coupled with significant advancements in imaging techniques, allow surgeons to better understand common and unique problems when presented with craniopagus twins (Browd et al., 2008). It has been shown that a surgeon's awareness of shared venous drainage is imperative for a successful separation of craniopagus twins (Bucholz et al., 1987), and Winston (1987) proposed that a classification system be developed based on the "deepest shared anatomical structure" (p 769). Despite huge advances in classification there are still significant areas missing. For example, there has yet to be a discussion of neural connections aside from their presence being correlated to higher incidence of neurological deficits following separation (Bucholz et al., 1987). Furthermore, Browd et al. (2008) are the first to document a classification system that takes complicated neural, vascular, and ventricular anatomy into account. However, there is still more research needed to understand how to classify shared neural tissue. The current classification systems led to Krista and Tatiana's shared circulatory

system being immediately recognized as a significant risk; nevertheless, classification failed to address their novel neural connection (Dominus, 2011a). Classification of neural connections may prove daunting, as variability in the connection is extreme; however, understanding the nature of neural connections may give surgeons the tool they need to remove the connection without causing neurological deficits. Surgical separation of craniopagus twins is a difficult task, perhaps the most formidable of all neurosurgery operations. Classification systems will help surgeons be aware of common problems, but surgeons must have a plan in place and be ready to tackle a host of obstacles once the twins are on the operating table.

Classification systems and imaging techniques aside, enhanced surgical and anesthetic techniques may be the most important factors contributing to the decreasing mortality rates seen in separated craniopagus twins (Browd et al., 2008; Bucholz et al., 1987; Girshin et al., 2006). Browd et al. (2008) indicate the importance of classifying surgical risk factors in order to determine mortality and the extent of surgery required. They then point to enhancing surgical techniques, stating that in the presence of a shared venous sinus, a staged separation is of great importance. In this series of surgeries – often six to eight – the surgeon's goal is to remove one of the twins from the shared sinus. This is done by slowly cutting bridging veins from the twin that will be removed from the sinus; the surgeon must not rush this process. Cutting too many bridging veins in one surgery may result in fatal bleeding (Browd et al., 2008). After each surgery, the venous system of the twin being removed will show increased blood pressure, resulting in angiogenesis, which is the formation of a new path towards their own deep venous drainage system (Browd et al., 2008). The process is repeated until one twin's drainage system completely bypasses the shared sinus, at which point the twins can be physically separated. Bucholz et al. (1987) first showed that this method provided the lowest mortality rate and best post-operative neural function. Huang et al. (2004) discuss a case in which a staged separation was not used. Serious complications arose, such as extreme blood loss and shock as a result of severe decrease in blood volume. These complications can be fatal, and they posed extreme challenges to the surgeons and anesthesiologists. By performing a staged operation, the surgeon avoids the most common complication associated with separating craniopagus twins: disastrous bleeding due to a shared venous drainage system (Browd et al., 2008).

Although staged separation was a key breakthrough in the surgical separation of craniopagus twins, carefully planned anesthetic management is crucial if the twins are to survive. Girshin et al. (2006) discuss the substantial benefits of using a staged separation and the challenges it poses to anesthesiologists. Namely, craniopagus twins must be put under and woken back up multiple times – each time with significant risk of complication. This is particularly troublesome since craniopagus twins often have other pulmonary and cardiac complications (Girshin et al., 2006). Additionally, if a shared circulatory system is present, medicine provided to one twin will affect the other twin while under anesthesia (Parameswari et al., 2010). These problems are significant but can be partially addressed by a strong pre-operative plan, the use of two separate anesthesia teams, and strong communication (Browd et al., 2008).

Another major complication may arise

in surgery that is often missed by current imaging techniques, and is not currently addressed in existent classification systems (Browd et al., 2008): shared neural tissue. Shared neural tissue significantly decreases the chances of optimal neural function after separation (Bucholz et al., 1987). Oddly, this risk factor has rarely been discussed in the literature. The enormous complexity of a neural connection is daunting; however, current research must begin to approach this issue and how it may be addressed in separation attempts. As craniopagus cases are rare in humans, this research may need to begin with animal models. In the case of Krista and Tatiana Hogan, separation posed too many risks. Not only do they share cerebral circulation (possibly a shared venous sinus) but also a complex neural connection, a bridge on which information can go from one girl to the other.

The Thalamic Bridge

Krista and Tatiana possess what their pediatric neurosurgeon Dr. Doug Cochrane has called a "thalamic bridge" (Dominus, 2011a). To appreciate the significance of this, a current understanding of the role of the thalamus is essential. Blumenfeld (2010) introduces the thalamus as "an important relay centre" (p 35). Pinel (2011) adds that "the most well understood thalamic nuclei are the sensory relay nuclei – nuclei that receive signals from sensory receptors, process them, and then transmit them to the appropriate areas of sensory cortex" (p 66). Dr. Cochrane describes the way in which Krista and Tatiana are unique: when sensory information travels to their thalamus it branches into two routes. Krista and Tatiana's thalami receive neural inputs, and process and relay them to the appropriate brain area. The inputs also cross the thalamic bridge to the other girl's thalamus (Dominus, 2011a). Krista and Tatiana share sensory information. To share every sensory modality passing through the thalamus allows them to not only feel what each feels but even see out of each other's eyes. A fine example of this emerges in an interview of the girls and their mother (Dominus, 2011b). Their mother covers Krista's eyes and holds a plush pony in front of Tatiana. She then asks Krista what she is holding and Krista replies, "pony." In the New York Times article, "Could Conjoined Twins Share a Mind?" Susan Dominus (2011a) describes an unpublished study performed by Dr. Cochrane when the girls were just two-years-old. While recording electroencephalography, or brain waves, from both girls, Dr. Cochrane covered Krista's eyes and flashed a light in front of Tatiana. Increased electrical activity from both girls' brain's visual areas was recorded. The test also worked when the girls switched roles (Dominus, 2011a). Surface electroencephalography is imprecise in its ability to localize where activity is happening but the fact that there is increased activity in both recordings is incredible.

Krista and Tatiana's ability to share sensory information is extraordinary; however, the thalamus does more than simply act as a relay centre. As stated by Pinel (2011), sensory relay nuclei are only a portion of the nuclei in the thalamus and describe merely one of its functions. Although the exact mechanisms are still unclear, the reciprocal connections from the thalamus to cerebral cortex are heavily involved in moderating consciousness. Blumenfeld (2010) poses a theoretical system, "the consciousness system," that regulates level of consciousness. He discusses three aspects of this system: alertness, attention, and awareness. These three aspects are all, in part, mediated by

nuclei in the thalamus such as the thalamic reticular nucleus, thought to preserve attention by gating information (Blumenfeld, 2010). Accordingly, it could be that as Krista and Tatiana age, it will become apparent that their levels of consciousness, which may affect spiritual understanding, self-actualization, and perception, stay very similar. As the thalamus is crucial in consciousness and sensory information processing, damage to it would be detrimental to Krista and Tatiana. This risk of damage, coupled with circulatory complications, led their physicians to conclude a separation would be too dangerous. Consequently, Krista and Tatiana are left with a connection that is both novel to documented research and exquisitely mysterious.

Continued Observation and Social Issues

Krista and Tatiana's thalamic bridge seems to connect one or more of their sensory modalities. It might also connect their levels of consciousness and perceptions of the world. Scientific exploration of this connection holds the potential to change modern society's view of individualism. Specifically, by observing Krista and Tatiana as they age, their unique concept of identity will become more apparent. Due to their connection, Krista and Tatiana will spend their entire life, as a pair, being extremely different from most people. Thus, as a pair, they paradoxically exemplify individualism. However, these girls face a challenge in individuality that has never been addressed. Although Krista and Tatiana have separate brains and personalities, they may share a common perception of their surroundings, which makes them rare even amongst craniopagus twins. Consequently, they embody the expression "seeing the world through someone else's eyes." The emotional repercussions of this ability are, at this time, a mystery; however, through observation, Krista and Tatiana's relationship may serve as an example of how empathy is expressed in its truest form. They will feel each other's pain and may struggle to differentiate their own sensations from their sister's sensations. Krista and Tatiana will struggle to find individual identity – as we know it. Their version of individual identity will, perhaps, be as half of a pair. They are described as using the word "I" to refer to the other twin; however, they are aware they are two separate people (Dominus, 2011a). Krista and Tatiana's conception of "I" may be different from everyone else's.

The twins' inability to differentiate between the personal and their sister may be one behavioural consequence of their neural bridge; in that, their inability to distinguish between their own sensations and those of their sister may confuse their sense of identity. Continued observation of Krista and Tatiana may result in the finding of other behavioural consequences caused by their thalamic bridge, which could further our understanding of the thalamus and individuality. Furthermore, the sharing of identity, levels of consciousness, and perhaps thoughts, has implications for our understanding of the philosophy of mind. If Krista and Tatiana share these complex notions of identity and thought – commonly associated with "the mind" as opposed to "the body" – it implies the mind may be shared through a physical bridge. The mind may not be separate from the body, but housed within it. Although the notion that the mind and brain are one is not new, Krista and Tatiana's case may further strengthen our understanding of how and where the mind is housed in the brain. Despite the

endless scientific advances Krista and Tatiana may provide, one must remember: they are individual girls, but, more importantly, an individual pair of girls. For us to understand how being part of a pair is more important than being an individual to one's identity, we must shift our perspective and eliminate our preconceived notions of individuality. Also, it is crucial that researchers be aware of the ethical complications coupled with pursuing research on the girls. Apart from possible developmental consequences, it may enhance the social stigma they already face. Additionally, the question of their voluntary consent must also be taken into account. Krista and Tatiana will face challenges arising from the possession of an ability no one in the world can fully understand. They simultaneously epitomize individuality as a pair while facing individual struggles never before seen.

Declaration of Conflicting Interests

The author declared they have no conflicts of interests with respect to their authorship or the publication of this article.

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