History of treatment of conjoined twins

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Abstract

This paper presents a history of the treatment of conjoined twins. The first mention of this malformation comes from the Neolithic period. Conjoined twins were depicted in mythologies of ancient peoples. The present paper focuses on the theories of formation of Siamese twins and attempts at their separation. Moreover, the history of treatment of conjoined twins in Poland is described.

Key words: conjoined twins, therapy, history

The oldest mention of the malformation defined as conjoined twins is believed to come from the Neolithic period. In Anatolia, the territory of contemporary southern Turkey, a 16.4 cm tall white marble figure dated at approximately 6500 BC was found with other excavations. It shows a two-headed goddess with explicitly visible twinning of the upper body part [1].

Another figure — the god Janus — with twinned upper parts of the body and two faces, one facing forward and the other backward, is found in ancient Roman mythology. Janus was a protector and guardian of the entry into and transition though gates of municipal walls and household doors, which was of extreme importance in those times as enemies often lurked outside the gates of towns-countries [2, 3]. Janus was also depicted as having four the faces, which may prove some influence of Roman myths on later Slavic beliefs, namely god Światowit (Svetovit) having four faces facing the four corners of the world.

In the beliefs of Eastern Slavs, there appears a three-headed God — Trzygłów — one of the most important gods for the Veleti. Trzygłów was worshipped mainly in Szczecin, Wolin and Brenna (Brandenburg) [4, 5]. The statues of Trzygłów had three faces looking in three directions. Its three pairs of eyes were embracing the sky, the earth, and the underground world. It is believed by some that the figure could have developed under the influence of Christianity and could have been a manifestation of the way the Slavs of the time imagined a Christian God in three persons.

One cannot however resist the impression that these unusual figures were modelled on people born with various congenital conjoined twins-like malformations [5, 6].

The literature available, including the Polish one, presents numerous descriptions of conjoined twins. Medieval chronicles, including Polish one, presents numerous descriptions of conjoined twins. Medieval chronicles, including Polish, often mention such phenomena in animals (“a two-headed calf”) as well as in people, however they are not trying to explain the condition, but merely for the sake of arousing the reader’s interest [7].

It can be assumed that such cases have always tempted doctors to attempt separation and aroused great interest among laymen; thus many pairs of conjoined twins gained popularity and many valuable and precise descriptions of their condition were provided.

Many researchers have attempted to explain the phenomenon of conjoined twins. The oldest preserved explanations come from Democritus and Empedocles (5th century BC) who, based on their research, concluded that conjoined twins appeared to be due to the improper composition and structure of spermatozoa. Aristotle (4th century BC) and later Galen (2nd century) accepted the above causes and added further external factors influencing the development of the malformation in question [8].

The Middle Ages did not produce any new theories as to the origins of conjoined twins, although in the 14th century an assumption was made that the possibility of its occurrence may be increased by a plague. Paré (17th century) in his book "Opera Ambrosii Parei Regis Primarii Parisiensis
Chirurgi” in the chapter entitled “De Monstris et Prodigiiis”, enumerated a variety of causes that may lead to the birth of monsters (both singular and twinned), including the following: an abundance of seed and overflowing matter (Gutmacher). The force and efficacy of imagination a fall suffered by a pregnant woman, the tone of the pregnant abdomen, hereditary diseases, or affects by any other accident, mixed the confusion and mingling together of the seed [9, 10].

In the 18th century, Lemery formulated his theory of fusion. He believed that conjoined twins were the effect of symphysis of two embryos due to various factors such as increased pressure. His theory was opposed by Winslow who claimed that conjoined twins came from one egg, which was to develop into separate embryos.

In the 19th and 20th centuries, despite considerable advances in medicine, especially in modern embryology, the causes of development of conjoined twins were not fully elucidated, and there remain differing theories describing the mechanism of their development [8].

Attempts to find the cause of this deformity have been the subject of numerous experiments on embryos of lower vertebrates. The studies of Born, Spemann, Schultz and Saint-Hilair demonstrated that many external factors such as shaking of the blastomere, merging of two gastrulas or narrowing of a blastula with a thread, a decrease in oxygen concentration and temperature, might lead to duplication of embryos [8].

Another theory, proposed by Willis and confirmed by Spencer, explains the malformation with secondary fusion of the developing embryos formed from a single zygote [11, 12].

The first documented attempt to separate conjoined twins was performed in Constantinople in the mid-10th century. A chronicler, Theophanes Continuatus, wrote that in 945 in Constantinople there appeared “a monster from Armenia”. Those were two boys from the same pregnancy, correctly developed, having normal external body parts, connected from the navel to the hypogastrium and facing each other (omphalopagus). At first, they were a curiosity, an attraction for citizens, but later they were treated as a bad omen and chased away from the city. They returned after some time during the reign of King Constantine. When one of the twins died, the most skilled doctors attempted to save the life of the other one and performed a surgical separation procedure. Unfortunately, three days later the other twin died as well. There was no mention of the use of anaesthesia or any analgesics in the original description of the procedure [13, 14].

Another thoroughly diagnosed case of pygopagus conjoined twins comes from England in the year 1100, when in Biddenden, Kent, two girls — Elizabeth and Mary Chulkhurst, were born united laterally at the buttocks (Fig. 1). They lived for 34 years; when one of them became ill and died, the other did not give consent for separation surgery, saying: “If we came to this world together — we will leave it together”. She died six hours later [15–17].

On September 10, 1495 in the village of Bierstadt near Worms in Germany, female craniopagus conjoined twins were born (Fig. 2). They were visited by Emperor Maximilian I who gave them 10 florins. In 1501 in Mainz, they were thoroughly examined and described by Sebastian Münster.
The scientist explained the case with the theory of imagination postulated by Paré: during pregnancy the mother of twins was gossiping in the street with another woman; the two accidentally clashed foreheads, and the woman became anxious. This imagination, he believed, led to the formation of symphysis of twins [16, 18, 19].

At the end of the 15th century, at the court of George III of Scotland, the ‘Scottish brothers’ – pygopagus conjoined twins, lived for 28 years [15].

Ambroise Paré in his work of 1682 entitled Opera Ambrosii Parei Regis Primarii et Parisiensis Chirurgi, in the chapter ‘De Monstris et Prodigis’, presented several cases of conjoined twins, which actually cover all currently encountered anatomical forms of this malformation (Fig. 3). One of the described cases is the abovementioned craniopagus malformation from Worms [10].

Another well-documented case is that of the ‘Hungarian sisters’ – pygopagus conjoined twins. Helen and Judith
were born in 1701 and were a public attraction. They lived for 22 years without an attempt at separation and died in a convent on February 8, 1723 [16, 20].

A similar case was that of Rose and Josephine Blazek born in 1878, who were a public attraction in Paris in 1891 (Fig. 4). They are also famous in teratology since one of them gave birth to a healthy child [16, 20].

According to the majority of reports, the first successful separation of omphalopagus conjoined twins was performed by König in 1689 (Fig. 5), who managed to separate them by gradual tightening of a thread ligature placed between the twins [21–26].

The term ‘Siamese twins’ comes from the most famous pair of xiphopagus conjoined twins — Chang and Eng Bunker, who were born in 1811 in Siam (now Thailand) (Fig. 6). At the age of 13, they were ‘discovered’ by an English merchant and at the age of 18, encouraged by an American sea captain, they moved to North America and assumed the surname of Bunker. Owing to their performances at the Barnum circus where they were shown as an unusual attraction — they made a fortune. Barnum used a clever trick — in each town they visited, he asked the local doctors for their opinion on the separation of brothers. Their statements printed in local newspapers evoked great interest among laymen, who themselves wanted to assess the possibility of successful separation. Naturally, Barnum himself was not interested in separation at all. Nevertheless, the examinations by different doctors give us thorough descriptions of the twins. At the age of 42, the Siamese brothers married sisters, of English origin, daughters of a pastor and settled down in North Carolina where they led the life of farmers. Unseparated, they lived for 63 years and had 22 children [16, 27].

Another famous pair of xiphopagus conjoined twins — Radica and Doodica — were separated at the age of 12 by von Doyen in Paris on February 9, 1902 (Fig. 7). The decision
as to separation was taken due to tuberculosis of one of the sisters (Doodica) and her quickly deteriorating general condition. During the separation of the liver and vessels, a severe haemorrhage occurred but the doctors managed to control it. Doodica died soon after the separation [27].

Another attempt at separation at the beginning of the 20th century concerned sternopagus conjoined twins (Maria and Rosaline) and was undertaken by Chapot-Prevost. Due to intraoperative complications, it ended with only a partial success (Fig. 8). Pneumothorax in Maria complicated by severe haemorrhage led to her death on the sixth postoperative day [27]. Unfortunately, the above reports do not specify the type of anaesthesia used during the procedure.

Numerous casuistic reports of Siamese twins were published in the Polish medical literature in the 19th century. The oldest mention of conjoined twins found by the author in the Polish specialist literature comes from the year 1837, from a doctoral thesis by Ludwik Przybyłko ‘Of incorrect position of organs’ written at the Jagiellonian University in Krakow. Przybyłko repeats after Meckel that “two-headed monsters with singular trunk are more common than one-headed ones with double trunk” [28].

The first mention of conjoined twins in the Polish Medical Review comes from the year 1887. Chamejdes, a physician from Zagórze, described and documented with his own drawings (Fig. 9) 2-week-old ischiopagus tripus conjoined twins [29].

In his paper, he aptly noticed that “the life of one of the two babies is endangered due to acute bronchitis, and the death of one of them would surely entail the death of the other”.

The following year, Browicz presented an anatomo-pathological description of this case as of a “double monster” [30].

In 1879, Eban, a district physician from Krzemieńczuk, member of the draft board in Gradziłsk in the Póltawska Province, noticed a malformation of “lower supernumerary monster limbs” in a conscript, a 21-year-old Cossack called Kondrat Kornuta (Fig. 10) [31]. A very detailed description of the anatomy and physiology of the defect, including the fact
that the autosite experienced pain from the parasite’s part of the body, is worth stressing. The editorial commentary by Jasiński includes the current classification of this malformation and, interestingly, its description and a comparison with similar cases reported in the literature (Rosenstiel, Winslow, Saint-Hilaire, Lancereaux, Virchow et al.) [32].

In the Medical Review of 1890, Patek — a doctor from the Stopnicki Province — presented a case of male twins conjoined with chests and stomachs down to the navel (thoracoomphalopagus). Since no movements of the foetus were detected for several days and the labour stopped, the foetuses were dismembered [33].

Another pair of Siamese twins was described in 1897. It was reported to be an extremely difficult labour case (Fig. 11) [34] and defined as a unique defect, which has never been described again. A similar case of conjoined twins “more than double” in people (the case of Galvagno from Catania) was described by Tur (Fig. 12), whereas Paré did the same in relation to animals (Fig. 13) [10, 20]. A triple conjunction was also mentioned by Guttmacher [9].
Janeczek presented obstetric difficulties in a 24-year-old woman from Książ Wielki in the Miechowski Province. Seizures with loss of consciousness developed during delivery, and, despite the administration of chloral hydrate and morphine, they did not subside. It should be stressed that analgesia or even chloroform anaesthesia (the author mentions that he “chloroformed” the parturient) was used during labour and two fragmented well-developed female foetuses, united at the navel area, were brought out [34].

More reports of conjoined twins were published in subsequent years. They are descriptions of specific obstetric complications. In 1935, Klein described a case of sternodymus [35].

In 1938, Korsyński presented a case of xypho-abdomino-pagus, which ended in the death of both the twins and their mother [36]. The next reports come from Kraków, where in 1946 dead thoracopagus conjoined twins were born and serious complications in the parturient were observed; in 1949, dead omphalopagus conjoined twins were spontaneously delivered [37, 38]. A case of ischiopagus tripus was presented in the report from the Department of Pathological Anatomy of 1950 [30]. The only Polish statistics taking into consideration the incidence of this malformation in our region is worth emphasising; it contains two cases out of 65,000 autopsies plus the report by Spitzer from Prague where only one case of ischiopagus was noted among 40,000 deliveries. Further reports come from 1956 (thoracopagus), 1962 (thoracoventropagus), 1965 (the only case of cephalothoracopagus described in the Polish literature), 1969 — two cases (thoracopagus, ischiopagus), 1980 (thoracopagus) and 1985 (thoracoamphalopagus) [39, 40–45].

The Polish reports have been devoted not only to casuistic descriptions of cases but have also presented the attempts at surgical treatment.

Information on the first separation attempt in Poland comes from 1969 [46]. A team led by Professor Poradowska in Warsaw conducted separation surgery in ischiopagus tetrapus Siamese twins. Both children died. The second attempt involved a parasite foetus (anterior thoracopagus parasite). Despite the doctors’ efforts, the child died shortly after separation from the parasite. Another surgery performed in the same clinic (asymmetrical incomplete posterior thoracopagus parasite) was successful [46]. In 1947, in Kielce, doctors conducted a successful separation surgery of children conjoined at the mid-abdomen [42]. Yet another surgery, the only one in Poland concerning cranioptagus parieto-occipitalis, ended in a complete failure [47]. The reports of successful separations of xiphamphalopagus and omphalopagus, performed in 1993 and 1995 respectively, are the only ones from outside our centre that have ended in success. Both pairs of twins are alive [48, 49].

References:

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