In a recent article in this journal, Hovorakova et al. reported on the anatomy of an anatomical specimen of a symmetrical cephalopagus conjoined twin from the collection of the Department of Teratology of the Institute of Experimental Medicine in Prague, Czech Republic [1]. Publication of case reports of complex congenital malformations is important for our understanding of teratogenesis and anatomy of these anomalies [2]. The report by Hovorakova et al., however, needs some additions and corrections.

1. Nomenclature

Although many different names can be found in literature (e.g., Cephalopagus, Cephalothoracopagus, Cephalothoracoileopagus, Syncephalus, Janus, Octopus, Janiceps, Monstre doubles sycéphaliens, Monstre à tête de Janus, Iniope, Synote, Déradelphe, Janiceps ateleus), the correct naming for this type of conjoined twin is cephalopagus. Cephalopagus conjoined twins (not to be confused with craniopagus conjoined twins) are fused from the top of the heads down to the umbilicus. There are two identical faces, on opposite sides of the large conjoined head, but in many cases (c. 75%) one of the two faces is rudimentary. Thorax and upper abdomen are always fused, the lower abdomen and pelvis never.

To avoid multiplicity of terms in literature in conjoined twins, only eight simple names should be used (Omphalopagus, thoracopagus, cephalopagus, ischiopagus, parapagus, craniopagus, rachipagus and pygopagus). To describe intermediates or aspecific cases, compound terms could be used.

As the thorax is always united in cephalopagus, 'cephalothoracopagus' is redundant and should not be used to describe the classic form [3]. The described case by Hovorakova et al. is also not an intermediate form, which should justify a compound term.

2. Rarity of cephalopagus

The authors stated, “There are seven main types of conjoined twins” and concluded several times that “Cephalothoracopagus Janiceps is the rarest form of conjoined twinning”. Both statements are incorrect.

Apart from the seven types of conjoined twins already mentioned by the authors in their article, there is an eighth recognised type to be distinguished: the rachipagus. This type of conjoined twinning is the rarest form and is only known from two published case reports [2,4,5,7]. Till 2005, rachipagus was only known from a case described by Bétouillères et al. in 1960 in a somewhat obscure French journal [5], but was brought to the attention of English speaking scientists by Spencer in 1995 [4]. At that moment many scholars still doubted the existence of rachipagus conjoined twins and considered the case of Bétouillères et al. a fabrication [6]. However, in 2005 Durin et al. reported on the first trimester diagnosis of a pair of symmetrical rachipagus conjoined twins [7]. Unfortunately this report showed some shortcomings, but the diagnosis of symmetrical rachipagus was crystal clear [2]. With the publication of the latter case report the existence of eight different types of conjoined twins was proven.

According to the incidence of occurrence, parapagus is the most common form of conjoined twinning. Spencer [8] studied 1200 cases of published conjoined twins and found 28% parapagus, followed by thoracopagus (19%), omphalopagus (18%), cephalopagus and ischiopagus (both with 11%), pygopagus (6%), craniopagus (5%) and rachipagus (2%). So, even when rachipagus is excluded, cephalopagus is not the rarest type of conjoined twinning, and is surpassed by pygopagus and craniopagus.

3. The gender of conjoined twins, especially of cephalopagus

The authors stated that: “For unknown reasons, 95% of conjoined twins are female. Based on these frequencies, the presented case of male cephalothoracopagus janiceps di-symmetros is the most extremely rare case of conjoined twinning” [1]. This is not in accordance with data in the literature. Although most conjoined twins are female, this is less than 95%. In a series of 312 cases of unselected conjoined twins, 61% was female [9]. In another small series of 23 unselected conjoined twins 48% was of the female gender [10].

The same figures can be found in cephalopagus. Spencer [11] studied 59 cases of typical cephalopagus of which 14 (24%) were male. Taruffi [12] collected 23 case histories of typical cephalopagus...
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4. Published cases of cephalopagus

In the discussion the authors stated that: “Concerning the presented di-symmetrical cephalothoracopagus janiceps, there are only few cases reported in the literature”. They cited only two case reports from 1990 to 2003. Unfortunately, they overlooked many published cases of di-symmetrical cephalopagus [13–37]. Beside these case reports of symmetrical cephalopagus conjoined twins, there are many more published reports of asymmetrical cephalopagus and parasitic cephalopagus. As is the case in many recent publications, well-described case reports prior to 1800 are neglected.

The oldest known published and illustrated case of a di-symmetrical cephalopagus was described by Edward Fenton in 1569 (Fig. 1) [38]. These twins were born in 1555 in Geneva and ‘had two faces as Janus . . . hadde’. The twins were delivered by embryotomy, and are for this reason depicted with two cut-off legs. Batman [39], Licetus [40], Aldrovandi [41] (Fig. 2) and Schott [30] also described and illustrated these conjoined twins.

5. Conclusion

Eight different types of conjoined twins can be distinguished; one is cephalopagus, which is fourth in rarity of occurrence. Between 17 and 24% of cephalopagus is of the male gender. More than 30 case reports of di-symmetrical cephalopagus can be traced in Western literature, the oldest published in 1569.

Conflict of interest

None.

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