Pygopagus

Pygopagus (Iliopagus): Two bodies joined at the pelvis, is one of the rare types of conjoined twins.

They are joined at the sacral area with sharing of terminus of spine, gastrointestinal system, genitourinary system and spinal cord to a variable extent. They represent a group of conjoints in which the separation of the embryonic axis in the caudal region was incomplete [1].

Pygopagus conjoined twins represent 6–19% of all the conjoined twins [2].

The reported incidence worldwide is estimated at 1:50,000 to 1:100,000, live births, with higher incidence of 1: 14,000 to 1: 25000, experienced in Asia and Africa [3].

Treatment

Twins with this configuration display symptoms and carry surgical risks during separation related to the extent of their connection which can include anorectal, genitourinary, vertebral, and neural structures. Neurophysiologic intraoperative monitoring (NIOM) for these cases has been discussed in the literature with variable utility.

The separation of conjoined twin is a unique challenge due to its complex anatomy and physiology. Although advancement in imaging and monitoring has improved the survival rates, separation can be successfully achieved only with meticulous planning and team work.

Case reports

Cromeens et al., present a case of pygopagus twins with fused spinal cords and imperforate anus where the use of NIOM significantly impacted surgical decision making in division of these critical structures [4].

Awasthi et al. present the case of one-and-half month-old male pygopagus conjoined twins, who were joined together dorsally in lower lumbar and sacral region and had spina bifida and shared a single thecal sac with combined weight of 6.14 kg. Spinal cord was separated at the level of the conus followed by duraplasty. They had uneventful recovery with normal 15 months follow-up [5].

Separation surgery of pygopagus asymmetrical conjoined twins sharing U-shaped spinal cord: case report and literature review [6].

A report describes the successful surgical separation of pygopagus twins who had a conjoined thecal sac and an epidermal cyst [7].

Multidetector computed tomography angiography for successful surgical separation [8].

1)
M. Winder, A. Law Separation of pygopagus conjoined twins: a New Zealand neurosurgical experience J Clin Neurosci, 13 (2006), pp. 968–975

2)
T. Hirokazu, I. Takayuki, H. Yoshinori, K. Kazunari, A. Akio, K. Keiji Separation surgery of Pygopagus asymmetrical conjoined twins sharing U shaped spinal cord: case report and literature review Childs Nerv Syst, 29 (2013), pp. 699–706
3)
R.M. Hoyle Surgical separation of conjoined pygopagus twins Surg Gynecol Obstet, 170 (1990), pp. 540–562
4)
Cromeens B, McKinney JL, Leonard J, Governale L, Brown J, Henry C, Levitt M, Wood R, Besner G, Islam MP. Pygopagus Conjoined Twins: A Neurophysiologic Intraoperative Monitoring Schema. J Clin Neurophysiol. 2016 Oct 1. PubMed PMID: 27749617.
5)
Awasthi R, Iyengar R, Rege S, Jain N. Surgical management of pygopagus conjoined twins with spinal bifida. Eur Spine J. 2015 Jan 21. [Epub ahead of print] PubMed PMID: 25599848.
6)
Hirokazu T, Takayuki I, Yoshinori H, Kazunari K, Akio A, Keiji K. Separation surgery of pygopagus asymmetrical conjoined twins sharing U-shaped spinal cord: case report and literature review. Childs Nerv Syst. 2013 Apr;29(4):699-706. doi: 10.1007/s00381-012-2007-2. Epub 2012 Dec 29. Review. PubMed PMID: 23274640.
7)
Chou YC, Peng HC, Chu CH, Tsai ST, Ling JC, Hsu YH, Chen SY, Harnod T. Successful separation of the conjoined thecal sac with an epidermal cyst in pygopagus twins. J Pediatr Surg. 2011 Sep;46(9):e25-7. doi: 10.1016/j.jpedsurg.2011.05.023. PubMed PMID: 21929972.
8)
Ohashi A, Tsuji S, Kuroyanagi Y, Kinoshita Y, Kaneko K, Mine K, Hamada Y, Inagaki T. Multidetector computed tomography angiography for successful surgical separation in pygopagus conjoined twins. Pediatr Int. 2012 Feb;54(1):150-2. doi: 10.1111/j.1442-200X.2011.03406.x. PubMed PMID: 22335328.