A Type I Split Cord Malformation is a Significant Pathomechanism of Spina Bifida Aperta in Neonates

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Abstract : Spina bifida (SB) is a well known spinal cord malformation which can result in serious neurological consequences in neonates. A new classification of SB based on the embryological pathomechanism of spinal dysraphism has been recently proposed. According to this classification, some SB may result from diastematomyelia in embryos and such SB is referred to as split cord malformations (SCM) and classified into Type I and II based on the presence and absence of an osseocartilaginous median septum, respectively. However, SCM are thought to be rare pathomechanisms of SB. We retrospectively reclassified 23 cases of apparent SB found in neonates (12 of SB aperta and 11 of SB occulta) according to the new classification. We found 3 cases of Type I SCM in the 23 newborns (13%) or 12 newborns with SB aperta (23%). All cases of Type I SCM presented SB aperta associated with Chiari II malformation with myelomeningocele. They all had paralysis in the lower extremities. Hypoplasia of the feet and clubfeet were seen in all patients while unilateral defects of the ribs were observed in a patient with thoracic SB. Found in 23%, Type I SCM therefore appears to be a significant pathomechanism of SB aperta in neonates.

Key words : **Spina bifida, Split cord malformations, Myelomeningocele, Chiari II malformation, Diastematomyelia, Notochord**