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Chiari 2 malformation and syringomyelia in a series of 236 patients with myelomeningocele

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A number of 239 consecutive patients with myelomeningocele are followed by the Centro Spina Bifida of the Ospedale Niguarda di Milano, with follow-up ranging from 2 to 30 years (mean 13.1 years). All patients were studied by cerebral and spinal MRI. *Chiari*-II malformation was evident in 236 patients (98.8%), and one or more *Syringomyielic Cysts* were present in 145 patients (63.7%). The *Chiari malformation* was responsible for some clinical symptoms (including mild swallowing disturbances) in 83 patients (35.1%). However, in most cases, symptoms tended to remain mild and spontaneously resolve with growth. Clinically relevant symptoms were seen just in 34 patients (14.4%). The syndrome occurred within 3 years of age in 18 patients, between 3 and 10 years in 7 patients, and below the 10th year in 9 patients. The clinical conditions tended to be more severe in younger patients, who generally experienced brain-stem syndromes, whereas the older subjects generally tended to present cervical cord syndromes.In all cases, the first treatment attempt consisted of CSF diversion or shunt revision. This resulted effective in 14 patients, while 20 required subsequent craniocervical decompression (CCD). Patients requiring CCD were either younger than 3 years or between 10-14 years of age, that means no patients between 3-10 years or below 14 years required CCD. Moreover, most CCD had to be performed in very young patients.Mortality consisted of 2 cases: a newborn with preoperative severe breathing anomalies, and a 7 year-old female, who had been operated in the neonatal period and deceased 7 years later because of shunt malfunction. All younger patients with initial brain-stem syndrome had stormy postoperative periods and required tracheostomy and digiunostomy, but ultimately achieved both respiratory and alimentary autonomy. Older patients with cervical syndromes usually had relatively smooth courses with slow progressive improvement.As to the S*yringomyielia*, this was symptomatic and/or progressive just in 6 of 145 cases (4.1%). Five of these 6 patients underwent CCD since the syringomyelia was thought as related to alteration of CSF dynamics at the Foraman Magnum, while one patient with lumbar progressive cyst was managed by marsupialisation. Three patients improved, two remained stable, and one kept on worsening in spite of subsequent multiple surgeries (cysto-peritoneal shunt, extended laminectomy etc.). The other 139 patients remained asymptomatic and with stable cysts throughout the follow-up.In conclusion, both Chiari-II malformation and syringomyelia are quite frequent in patients with myelomeningocele, but they require treatment just in a minority of cases. Both conditions present peculiarities, which make them quite different from analogous lesions in patients without myelomeningocele.