

Prospective, Interdisciplinary Follow-up of Children with Prenatally Diagnosed Giant Omphalocele: Short-term Neurodevelopmental Outcome

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PURPOSE: The objective of this study is to determine the short-term neurodevelopmental outcome in infants with giant omphalocele (GO).

MATERIALS AND METHODS: Between January 2002 and December 2007, 31 consecutive infants with GO received prenatal and postnatal care at our institution. Overall survival was 81% (25/31). Twenty (80%) of the survivors were enrolled in a prospective interdisciplinary follow-up program. Fifteen were at least 6 months of age and received detailed neurodevelopmental evaluation using the Bayley Scales of Infant Development II (BSID-II [before 2006, n = 3]) or BSID-III (after 2006, n = 12). Scores were grouped as average, mildly delayed, and severely delayed by SD intervals (115-85, 71-84, <70). Scores were considered mixed if cognitive or language skills were in different ranges.

RESULTS: Median age at evaluation was 12 months (range, 6-26 months). Average, mildly delayed, and severely delayed scores for cognitive and language skills were found in 6 (40%), 2 (13%), and 6 (40%), respectively. One child had mixed scores (severely delayed for cognitive and mildly delayed for language skills). Motor scores were normal, mildly delayed, and severely delayed in 6 (40%), 2 (13%), and 7 (47%), respectively. The neuromuscular examination was abnormal in 8 patients (62%). Five (33%) scored within the average range, whereas 6 (40%) demonstrated severe delays for cognitive, language, and motor outcome. Of the 6 children with severe delays, 2 (13% of total) have autism, 4 required tracheostomy, and 1 was diagnosed with Williams syndrome.

CONCLUSIONS: The presence of GO is associated with deficits in developmental achievements in most of the affected infants ranging from mild to profound delays. These findings underscore the importance of early and standardized neurodevelopmental evaluation throughout childhood for all survivors with GO. Larger studies are warranted for risk factor stratification.

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