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CORPUS CALLOSUM AGENESIS AND DEVELOPMENT

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The authors analysed clinical files of 3738 children followed at the Child Development Department, Santa Maria University Hospital - Lisbon- between January 1990 and March 2003.
29 cases of corpus callosum agenesis were identified (18 male ; 11 female), aged between 7 days and 4 years at first observation (mean 11 months).
Psychomotor development was assessed with the Denver II Developmental Screening Test or the Ruth Griffiths Mental Developmental Scales. Developmental impairment was considered when GQ was below 80 in the Griffiths Scale and below the 25th centile in the Denver Scale.
Diagnosis was prenatal in 3 cases and postnatal in the remainder (five in the neonatal period).
Corpus callosum agenesis was total in 14 children (48%) and partial in 15 (52%); 23 children had other abnormalities (CNS or others).
In 6 cases (21%) genetic etiology was identified: 4 chromossomal abnormalities; 2 genetic syndromes.
In 3 cases consanguinity existed ; 2 other cases had a family history of corpus callosum agenesis (a sibling in both cases).
Development was assessed in 18 children using the Griffiths Mental Developmental Scales: 12 had developmental impairment (GQ below 80), 4 had a normal development (GQ between 80 and 100) and 2 were above average (GQ over 100). In 11 cases the Denver Scale was used: 5 were below the 25th centile and 6 above.
Comments:
Corpus callosum agenesis does not necessarily imply developmental impairment. Some children in our sample had a normal or above average development. Impairment was usually present when other malformations co-existed.