chological well-being and feelings of inadequacy in relation to their parents and family. The Shwachman score of the sick child was not related to the level of self-esteem. Conclusion: When CF is present among siblings, girls seem to carry more of the family pain than boys, a finding that calls for an increased awareness of the girls' situation by members of care teams.

性别影响了囊性纤维化患儿及其健康同胞的自我评价

目的：评价在囊性纤维化（CF）患儿及其健康同胞的自尊感是否与健康对照组不同，以及在同胞对内和同胞对间的自尊感是否不同。方法：所有 6～14 岁的瑞典 CF 患儿及其相同年龄范围的健康同胞（n = 65）受邀参加研究，55 个同胞对及其父母参加了研究。通过“我认为、我是”自我评价调查问卷对 5 个方面的儿童的自我意识进行了评价：身体特征、技能和才能、心理健康、与父母和家庭的关系以及与其他人的关系。疾病的严重性通过 Shwachman 临床评价体系的方法来评估。结果：尽管在一般水平上组间的自我评价无差异，但是健康女童以及那些有 CF 的女童与对照组的女童相比，在“心理健康”和“与父母和家庭的关系”等方面的评分较低。对性别组合（患病女童/健康男童、患病女童/健康女童、患病男童/健康男童、患病男童/健康女童）的比较提示女童对其心理健康的感觉较差且有与其父母和家庭有关的不满足感。患儿的 Shwachman 评分与自尊感的水平无关。结论：当在同胞中存在 CF 时，女童似乎比男童感受到更多的家庭痛苦，这一结果要求保健人员增加对女童状况的关注。

0274. Ages and Stages Questionnaire used to measure cognitive deficit in children born extremely preterm

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Aim: To validate the Ages and Stages Questionnaire (ASQ) and to measure average cognitive deficit in children born extremely preterm. Methods: Parents of 30 term children aged 36－42 mo completed the ASQ and the children underwent the Wechsler Preschool and Primary Scales of Intelligence-Revised. In a second study, the ASQ was obtained in 22 children born extremely preterm and 19 term children at the age of 35－44 mo. Results: The overall ASQ score correlated significantly with IQ (p = 0.007). The children born extremely preterm had an ASQ score of －1.06 SD below the score of the term children (p = 0.048). Conclusion: The ASQ identified a developmental deficit of the expected magnitude.

ASQ 用于测定极早产儿的认知障碍

目的：验证“年龄发育进程问卷”(ASQ)并测定极早产儿的平均认知障碍水平。方法：30 名月龄在 36～42 个月的足月儿的父母完成了 ASQ 并且这些儿童进行了修订的 Wechsler 学龄前和小学智力量表》测定，在第 2 次研究时对 22 名极早产儿和 19 名足月儿在月龄 35～44 个月时获取 ASQ。结果：总体 ASQ 评分与 IQ 显著相关（P = 0.007），极早产儿的 ASQ 评分比足月儿的 ASQ 评分低 1.06 倍的标准差（P = 0.048）。结论：ASQ 识别出了预期数量的有认知功能发育障碍的新生儿。

0275. Neurodevelopmental outcome and haematological course of a long-time survivor with homozygous alpha-thalassaemia: Case report and review of the literature

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Aim: Homozygous α－thalassaemia, also called haemoglobin (Hb) Bart's hydrops fetalis, has been thought to be a lethal condition. Due to prenatal diagnosis and intrauterine blood transfusions, a few patients with Hb Bart's hydrops fetalis have survived. This fact raises the urgent questions of clinical management and appropriate follow-up of these patients, both of which are addressed in this article. Methods: We report on a 6.5－y-old patient with homozygous α－thalassaemia and review the literature of 13 other survivors published to date. Transfusion requirements were evaluated and the rate of liver iron accumulation was assessed by biomagnetic liver susceptometry before and after institution of iron-chelating therapy. Psychometric evaluation was carried out using Munich's Functional Development Test, the Columbia Mental Maturity Scale, the Kaufman Assessment Battery for Children, and the Peabody Picture Vocabulary Test. Results: Our patient had significant delay of psychomotor development. Psychometric evaluation at the age of 5 y revealed an IQ of 85 and an intellectual level of a 4－ y-old child. Early tissue iron overload was seen, but a negative iron balance was achieved after institution of desferrioxamine treatment at dosages used for β－thalassaemia. Conclusion: Ho-