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MINI REVIEW

Tips and hints for the transition: What adult hepatologists should know when accept teens with a pediatric hepatobiliary disease

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Summary The number of children with chronic hepatobiliary disease surviving into adulthood is more and more increasing, but no established model of transition does exist in this category of patients. Here, we summarize medical problems expected at the time of their transition, and any impacts on morbidity and mortality in adulthood. Information provided would turn useful to adult hepatologists and practitioners responsible for ensuring continuity of care for young adults affected by diseases they are not usually accustomed to.

Introduction

The effects of a chronic illness on adolescents can significantly affect both biological and psychological development, sometimes deteriorating also the cognitive and social relationships [1]. Transition of patients with paediatric onset chronic diseases to a service for adults, if based simply on their chronological age without adequate parties' preparation, is therefore a challenging condition. This is in fact complicated by the intrinsic still ongoing adolescence process and the usual scarce information of adults practitioners on the long-term outcome of these sometimes ''unusual'' diseases as well.

For that reason, paediatricians are often encouraged by specific sub-specialties scientific associations to develop strategies for overcoming these barriers putting emphasis on patients' self-management, improvement of knowledge about their own condition, and supporting the decision-making process. Different models of transition for several paediatric onset chronic diseases (diabetes mellitus, cystic fibrosis, rheumatologic diseases, nephrology) have already been proposed. On the contrary, there is not yet a

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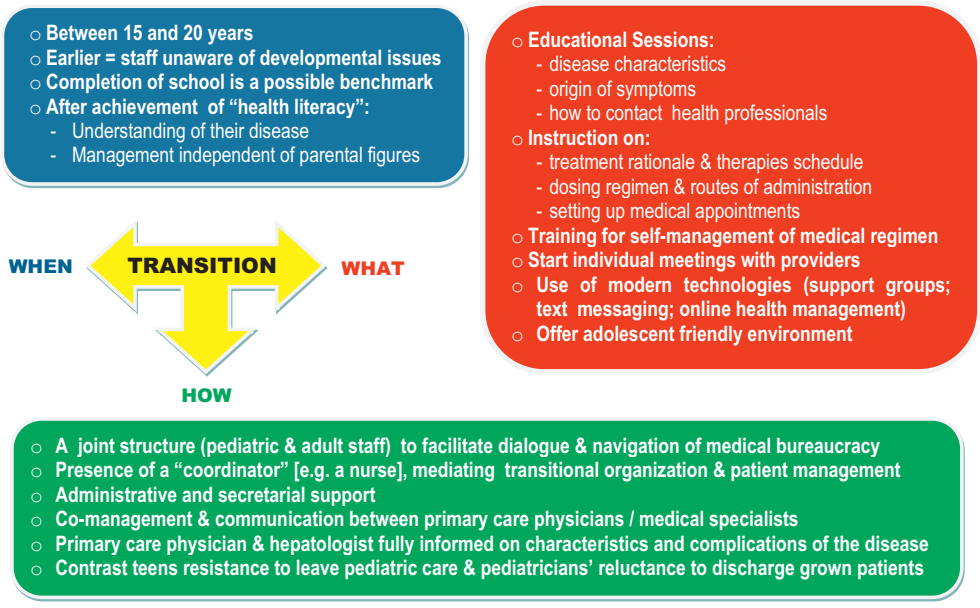

Figure 1 Proposed general tips for an effective transition program of a pediatric onset chronic disease [2,20].

unique and well-structured transition program for paediatric patients with chronic liver disease, which — in the last decades — have significantly increased their survival rates and quality of life as well. Pending sound societary ad hoc guidelines, we have recently reviewed a number of features that will probably need a further focus [2]. Fig. 1 summarizes schematically the most important aspects of the review.

The present article in particular takes into account a number of papers reporting long-term outcomes available for paediatric onset chronic liver disease (Table 1). Problems expected at the time of transition, and any impacts on morbidity and mortality in adulthood are outlined. We intend to provide useful evidence based medicine information to adult hepatologists and practitioners responsible for ensuring continuity of care for young adults affected by diseases they are not usually accustomed to.

Biliary atresia

Patients with biliary atresia can survive without Liver Transplantation (LTx) > 20 years after Kasai intervention performed during early childhood (survival at 5, 10 and 15 years: 63%, 54% and 44%, respectively). However, more than half of the patients ultimately are subjected to hepatobiliary disease related long-term complications, including hypersplenism and major episodes of cholangitis [3]. Hepatopulmonary syndrome and portopulmonary hypertension may also be a problem.

The ever increasing number of patients with biliary atresia surviving $\geq$ 20 years after Kasai operation requires permanent meticulous care to ensure a decent quality of life, allowing for example, also safe and well controlled pregnancies [4].

Approximately 80% of patients, who experienced cholangitis, subsequently develop portal hypertension, with possible gastrointestinal bleeding, and ultimately liver failure. Hepatocellular carcinoma degeneration is also possible.

For these reasons, it is important to consider the proper timing of liver transplantation, in order to avoid the risk of mortality related to long-term complications after Kasai vs the intrinsic risks of a long-term immune suppressive therapy.

Autoimmune liver diseases

The term autoimmune liver disease encompasses a spectrum of related diseases with variable presentation. Rapid progression in the case of delayed or ineffective conventional therapy with immunosuppressants may ultimately require the need for LTx [5]. In particular, autoimmune hepatitis in children is associated with a significant percentage of survival lower than the general population. The detection of cirrhosis on initial liver biopsy does not seem to have an impact on long-term survival.

Despite the high and prolonged pre-LTx immunosuppression, post-transplantation results for autoimmune hepatitis are similar to patients undergoing LTx for other indications, without significant increase of infectious, metabolic, and neoplastic complications [6].

A form of de novo autoimmune hepatitis interestingly affects children transplanted for non-autoimmune conditions and requires the same treatment schedule used for classical autoimmune hepatitis.

Primary sclerosing cholangitis is increasingly diagnosed in children and adolescents, due to the greater spread of magnetic resonance cholangiography, but its long-term prognosis remains uncertain. Its overlap with autoimmune hepatitis (overlap syndrome) can respond to the same treatment used for autoimmune hepatitis as regards to the parenchymal inflammation, but in about 50% of cases, the biliary ducts disease may progress, leading to a worse prognosis and a greater need for liver transplantation. Although drug therapy (ursodeoxycholic acid [UDCA]) appears to improve symptoms and liver tests, there is no influence on long-term