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SPECIAL ARTICLE

Liver and intestinal transplant in the paediatric population☆

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Abstract Our organizational model allows an annual running of 1000 liver transplants. Paediatric liver transplantation constitutes 5% of such activity and provides, in children with severe, progressive and irreversible liver disease, a 1-year survival of 90% and more than 80% after 15 years of follow-up. The main indication is biliary atresia followed by metabolic liver disease and acute liver failure. Around half of the procedures are performed in children under 2 years and 25–30% in the first year of life.

The waiting list remains at around 35 patients, with an average of 100 patients enrolled annually and 60 of them finally transplanted after an average of 136.3 days on the waiting list. The prioritization of the candidates uses the PELD as an objective tool for decision-making. However, the progressive ageing of donors, with a profile increasingly different from the requirements of the paediatric patients included in the waiting list, requires strategies such as living donor liver transplantation and the split liver transplantation, to increase the probability of transplant while reducing both time and mortality on the waiting list at the same time.

Paediatric intestinal transplantation registers a low indication but involves strict requirements that outline a very uncommon donor in our country which, together with the absence of alternatives that outweigh the impact of these difficulties, penalizes the chances of transplant for these patients.
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PALABRAS CLAVE
Trasplante hepático pediátrico;
Trasplante intestinal;
Trasplante multivisceral

El trasplante hepático e intestinal en la población pediátrica

Resumen Nuestro modelo organizativo posibilita la realización anual de 1.000 trasplantes hepáticos, de los cuales el trasplante hepático pediátrico constituye el 5% y proporciona, en niños con hepatopatía grave, progresiva e irreversible, una supervivencia del 90% al año y superior al 80% a los 15 años de seguimiento. La principal indicación es la atresia de vías biliares

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seguida de hepatopatías metabólicas e insuficiencia hepática aguda, realizándose el 50% de los procedimientos en menores de 2 años y el 25–30% en el primer año de vida.

La lista de espera se mantiene en torno a los 35 pacientes, con un promedio de 100 pacientes incluidos anualmente y 60 trasplantados tras un tiempo medio de espera de 136,3 días. La priorización de los candidatos utiliza el PELD como herramienta objetiva de apoyo en la toma de decisiones. No obstante, el progresivo envejecimiento de los donantes, con un perfil cada vez más alejado de los requerimientos de los pacientes infantiles incluidos en lista de espera, precisa impulsar estrategias como el trasplante hepático de donante vivo y la modalidad split, para incrementar las probabilidades de trasplante reduciendo la mortalidad en lista de espera y el tiempo de permanencia en la misma.

El trasplante intestinal/multivisceral pediátrico registra una baja indicación pero conlleva unos requisitos que perfilan un donante muy infrecuente en nuestro país, lo que, unido a la ausencia de alternativas que contrarresten el impacto negativo de estas dificultades, lastra las probabilidades de trasplante de estos pacientes.
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The Spanish organizational structure for organ donation and transplantation has allowed Spain to achieve the highest rate of donation worldwide continuously since the early 1990s, exceeding 35 donors per million population (pmp).[1] As a direct consequence, between 1000 and 1100 liver transplants can be performed in Spain each year, reaching a rate of 23 procedures pmp, one of the highest in the world.[2,3] Among these procedures, paediatric liver transplantation accounts for 5% of the activity nationwide, with a mean of 60 transplants per year.

We ought to note that paediatric liver transplantation is an infrequent procedure due to both the low incidence of the diseases for which it is indicated and the reduced availability of donors that are suitable matches for the recipients on the waiting list, as the donor pool has been ageing on a sustained basis. This becomes evident when we analyze the increase in the mean age of donors in our country, which has practically doubled from 36 to 60 years over the past two decades. In this regard, data from 2014 revealed that only 1.5% of registered donors in Spain were younger than 15 years.[1] The comorbidities present in current donors combined with the ageing trend pose challenges to donor–recipient matching, and this has the most negative impact on the paediatric population.

In order to alleviate the negative repercussions that result from the particularities of this procedure, the Consejo Interterritorial del Sistema Nacional de Salud (Interterritorial Council of the National Health System) considers the sites that perform it reference centres, departments and units (Centros, Servicios y Unidades de Referencia [CSURs]). The purpose of this designation is to guarantee equity of access, regardless of the place of residence, and quality services to individuals with rare and complex diseases that require highly specialized care, which in turn necessitates the concentration of patients and health care resources.

Indications for transplantation and waiting list management

Paediatric liver transplantation is indicated for severe liver disease for which there is no other effective treatment when it has been determined that performing the transplant can improve survival and quality of life compared to not performing it, an assessment that requires a precise diagnosis.[4,5]

Good outcomes in the post-transplant survival of children have led to a shift in the approach to the management of severe paediatric liver disease.[6,7] In chronic cases, the risk of death due to liver disease is not the only determinant in the decision to transplant or not, and another factor under consideration is the severe impairment of quality of life, due to a pathology for which there is no other treatment of comparable efficacy and sufficiently developed, that would justify the risk of early death associated with transplantation.

Thus, it is essential that the risk of death be assessed in order to determine the optimal timing for transplantation, anticipating the development of predictable severe complications, and also to assign degrees of priority to the patients on the waiting list managed by each site.

Activity data show that around 5% of patients on the waiting list for a liver transplant belong to the paediatric age group, a percentage that is similar to the one found in the total population of transplanted patients, which shows that the probability of receiving a transplant is maintained in this age group in spite of its particular challenges.

The childhood diseases that may be treated with a liver transplant mainly include cholestasis, metabolic disorders, cirrhosis and severe acute liver failure. The main indication for liver transplantation in children is biliary atresia, which accounts for 25% of the patients in the waiting lists in Spain. Metabolic liver disease accounts for the second highest number of cases included in the waiting lists, followed by acute liver failure, accounting for 13% and 10% of transplant candidates, respectively (Fig. 1).

Similarly, if we analyze the transplantations performed in 2014 (Fig. 1), biliary atresia remained the most frequent disease in children that underwent liver transplantation in Spain (27%), although acute liver failure had become the second most frequent indication (18%), surpassing metabolic diseases (8%). This change was promoted by the prioritization of those cases, which increased their chances of receiving a transplant. Approximately 3–4% of transplants

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