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The start of the transplant journey: Referral for pediatric solid organ transplantation

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Abstract

The focus of the majority of the psychosocial transplant literature is on post-transplant outcomes but the transplant journey starts much earlier than this, at the point when transplantation is first considered and a referral for transplant evaluation is made. In this review we cover information regarding the meaning of the referral process for solid organ transplantation. We discuss various factors of the referral for transplantation including the impact of referral on the pediatric patient and the family, potential expectations and misconceptions held by pediatric patients and parents, the role of health literacy, decision making factors, and the informational needs of pediatric patients and parents. We elucidate steps that providers can take to enhance transplant referral and provide suggestions for much needed research within this area.

Keywords

pediatric transplantation; abdominal; cardiothoracic; psychosocial adjustment; referral

Introduction

Solid organ transplantation is a widely respected method for the management of various disease states refractory to medical treatment [1–6]. The number of pediatric solid organ transplants performed has continued to increase with approximately 43,000 transplants completed in children aged 0–17 years in the United States between 1988 and 2013 (Figure 1; Based on OPTN data as of October 21, 2013). Improvements in graft and patient survival over the last decades [1, 5] have made solid organ transplantation a viable and at times

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preferred treatment alternative for infants, children, and adolescents. While in the past solid organ transplantation was strictly a lifesaving procedure, now transplantation is not only life extending but also thought of as potentially life enhancing [2, 7, 8]. As pediatric solid organ transplantation has become a more established treatment avenue, referral for transplantation has become an increasingly important step in the transplant process.

Medical considerations for the referral to transplant evaluation vary by disease state. However, parents of children who are at the end stage of their disease [2, 3, 8] be it renal, hepatic, intestinal, cardiac, or respiratory have been provided with some information regarding the potential for transplantation. Similarly families of patients with acute presentations of disease have typically been approached about transplantation early on as a primary treatment option. At this time, however, little data exists in the pediatric literature regarding the best approach to introduce solid organ transplantation as a treatment alternative [2, 7]. In addition, information is scarce regarding parental and pediatric patients' reactions to referral for transplantation [9–11] and the potential impact of the referral process on the subsequent evaluation. Given the implications that referral for solid organ transplantation can have for a family and patient, we believe that a discussion of the factors that could influence this process would be helpful with specific attention devoted to the aspects that affect the rest of the transplant journey.

Meaning of transplant referral for children and their families

It is estimated that before their 18th birthday 10% of children will be diagnosed with a chronic illness (defined as a condition which has been present or is likely to be present for at least 3 months and is not (yet) curable [12]) that is severe enough to significantly limit their daily life and necessitate extended care and supervision. [13] Whilst there is an extensive literature on the psychological, emotional and social impact of chronic illness for children and families (e.g., cancer and diabetes) and a growing literature of palliative and end of life care in pediatrics, far less is known about the impact of end-stage organ failure necessitating transplantation.

The disease course resulting in consideration for transplantation varies within and between organ types as well as by disease presentation. For some patients the referral for transplantation is viewed as a potential new beginning while for others the discussion of transplantation signals the end of a long series of attempts to manage and improve a progressive and chronic medical condition. Patients and parents are often unsure of what transplant may hold/provide and uncertain of the transplant process. Consequently, the impact on the child and family of reaching the point where a transplant may be necessary will vary based on multiple factors including individual and family differences in social and psychological functioning, illness course and presentation, previous treatment course, and known transplantation outcomes. Although these experiences vary widely, one unifying factor is that referral for transplant typically indicates a significant shift in treatment approach. Hence, a non-categorical approach [14] to understanding the psychosocial impact of end-stage organ disease and transplant referral is a useful technique to understanding the meaning of transplantation referral.

Impact of end-stage organ failure on children and adolescents

Children are referred for transplant for a number of congenital (e.g. complex congenital heart disease, biliary atresia, prune-belly syndrome) and metabolic (e.g., Maple Syrup Urine Disease, Hurler's syndrome, cystinosis) conditions which may be associated with developmental and cognitive delay, neurocognitive deficits, and central nervous system damage. [15–19] Furthermore, there is evidence that children and teenagers with long-term conditions who develop end-stage disease are at risk for cognitive deficits, mood disorders, low self-esteem, poor peer relationships, behavior problems and adjustment difficulties, [20–25] all of which can have an impact on their ability and willingness to entertain and/or discuss transplantation as a treatment option. Some children may be considered for transplant due to conditions which are secondary to previous illnesses or treatments (e.g. bronchiolitis obliterans after stem-cell transplantation and cardiomyopathy as a result of chemotherapy) that may involve psychosocial and cognitive sequelae. This group of young people will already have had to cope with the psychological and physical impact of a potentially life-threatening illness and treatment regimen, and the subsequent referral for solid organ transplantation may be very traumatic.

Children who develop sudden onset acute organ failure have to cope with the other end of the spectrum. Having been previously healthy and typically normally developing with few previous restrictions, these children face situations requiring urgent and often invasive medical interventions that drastically alter their quality of life [26, 27]. Unlike those who have a long history of illness, these patients frequently have no experience of illness, hospitals, or treatment regimens and may also be too ill to effectively participate in decision-making. These patients are typically less well versed in medical terminology and experience a very sharp learning curve. In addition, these patients may have to quickly readjust expectations of their future and proceed with treatment that they may not fully understand.

The need to assess children's psychological adjustment prior to transplant is now being recognized, however, this is rarely undertaken in the referring center. The mental health status of the patient is of particular importance as psychological functioning of the child/adolescent has an impact on post-transplant outcomes, including adherence to medical care [28]. Increasingly psychological assessment data are being collected at the transplant center. Reported data are typically based on small single site samples, although it is now more common for transplant registries to include some developmental, cognitive and/or psychosocial parameters even if most of these are collected post-transplantation [4, 5]. One of the potentially confounding factors affecting pre-transplant data is that the data are collected during transplant evaluation. Pre-transplant evaluation is a stressful time when potential transplantation has become a true possibility for patients and families. Such times of heightened stress are not ideal for effectively capturing a baseline of functioning, but are nevertheless helpful in potentially identifying mental health, academic, and social support concerns that could have an impact on the rest of the transplant evaluation and overall transplant process.

Impact on the family

There is a body of evidence documenting the stresses faced by parents of children with end-stage kidney disease and to a lesser extent parents of children with cystic fibrosis facing the need for transplantation [7, 9, 29]. High levels of anxiety, depression and stress are reported among parents of these children [30]. Recent studies have identified PTSD symptoms among parents of solid organ transplant recipients and suggest a higher prevalence of these symptoms in the parents of transplant patients than among other groups of parents coping with chronic medical conditions (e.g., HIV) [31, 32]. In general, however, there are few data concerning parental adjustment at the time when transplant referral is being considered. In particular, there is a dearth of evidence related to patient or parent adjustment in situations where there is a rapid onset of organ failure.

It is acknowledged that children with chronic or end-stage disease frequently have complex developmental and emotional needs which place additional demands on the family [33]. However, it is also increasingly evident that it is not only characteristics of the disease which determine how a family adjusts to a child's chronic illness, but also how families perceive and cope with the illness. Adjustment of the child and family is further influenced by the medical processes that occur around them and medical information available to them. For example, while guidelines for transplant referral and listing are pretty standard in some disease specific groups [34, 35] there appears to be less consensus in some areas (e.g., intestinal failure and short gut syndrome) [3] about the right time to refer and list. This in itself has clear psychological implications for children and their families as they may receive conflicting messages from medical providers as well as have misconceptions and inaccurate expectations regarding the transplant process.

Issues such as the wider impact on the family of transplant referral (e.g. in terms of arrangements for siblings and practicalities related to travel, employment and finances), the prospect of leaving one health care team, the need to develop relationships with a different team, and coping with the uncertainties associated with transplant referral have not been investigated in families prior to their arrival at the transplant center. Each of these issues carries significant ramifications for the patient and family and should be considered equally important components of the referral and subsequent transplant process.

Impact of the timing of transplant referral

One of the tasks of a referring center is to balance the medical and psychological considerations when introducing the idea of transplantation [36]. An early referral provides children and families with more time to assimilate a large volume of complex information, reach a stage of emotional acceptance of the key issues, and make an informed decision regarding treatment options. It also gives the referring center more opportunity and time to prepare and support the family while giving the transplant center an opportunity to build relationships with the child and family [36]. Conversely referring too early may cause unnecessary psychological distress for families already facing challenging medical circumstances [36].

A fine balance has to be reached such that referrals are made at a time when the medical status of the child can be optimized. If a referral is made too late a child may become too sick and a family too distressed, thus compromising optimal care and potentially precluding transplantation as an option. Referring too late for transplantation could also lead to the patient being urgently listed, providing insufficient time for the child and family to discuss the various options, assimilate the information and make a truly informed decision. In some cases where deceased donor organ transplantation is the only possibility a late referral may mean that a child dies before a donor organ becomes available. Between 1995 and 2013 3,576 children died waiting for an organ in the United States (based on OPTN data as of October 21, 2013).

Expectations and misconceptions about transplant

How families respond to learning that their child may need a transplant will vary in part by the type of transplant needed. While being faced with a possible transplant is undeniably stressful, children and families presented with a type of transplant that has been done many times with high long-term survival rates (e.g., kidney and liver transplantation) may be less stressed and more optimistic about prognosis compared to children/families facing a type of transplant that is relatively rare, only recently performed, and where long-term survival data are limited or poor (e.g., intestinal and lung transplantation).

Regardless of the type of transplant, it is important to clarify the child/family's expectations for transplant, and correct any misconceptions that they may have. For example, families need to understand that transplantation is not a cure, but rather a treatment option that extends life. Some families expect that transplantation will end a long road of persistent medical intervention and are disappointed to learn that transplantation is a different, albeit hopefully better, road with ongoing need for medical management. Families are also surprised by the "longevity" of transplanted organs. For most types of transplant, the new organ is not expected to last throughout the remainder of the child's life; thus the child may face the need for another transplant in the future. For patients who are headed toward transplant because of a chronic illness, transplant can be conceptualized as exchanging one chronic illness for another [37, 38]. For patients with acute presentations, the idea of "saving and extending" life is of particular importance. Patients and families should understand that receiving a transplant results in the need for life-long medical care including ongoing medical visits, procedures such as blood draws and biopsies, and taking daily medication.

Families may also expect that except for the surgical risks and need for ongoing medical management transplantation carries few other considerations. Emphasis should be placed on educating families that there are long-term risks and potential complications beyond survival, including the risk of infection due to being immunocompromised, the risk of rejection, and other medical issues such as post-transplant lymphoproliferative disorder (PTLD), or other malignancies [1, 39–41]. Specific organ transplants may have additional risks; for example children undergoing heart transplantation are at risk for developing vasculopathy of the coronary arteries, which could result in the need for a second transplant [1]. In addition, many children who have undergone nonrenal solid organ transplantation may eventually require kidney transplantation [42].

Expectations and misconceptions of donation

Expectations and misconceptions of organ donation should also be touched upon. At times parents of children being referred for deceased donor organ transplantation may conceptualize the situation as “someone has to die for my child to live”. Similarly, older children and adolescents may also experience concern regarding the donor as well as worry about having someone else’s organ inside their body. Other potential beliefs such as that a transplanted organ may alter a recipients personality, tastes, or cognitive processes could also be harbored by both children and their parents. Children and parents should be educated about the deceased donor process and their misconceptions reframed to avoid guilt, stress, and/or fear of receiving an organ.

Additional expectations and potential misconceptions need to be considered for children/families in which transplant includes the possibility of receiving an organ from a living donor. Families who are presented with the option of living organ donation may believe that this option is somehow easier and preferred, as the donor is known to the child/family, and in many cases, is the parent, relative or sibling of the child needing a transplant. Families may think that once a living donor volunteers and is a “match” medically, that transplant can proceed, being scheduled at a time that is convenient for the donor and recipient. Children/families need to be educated that in addition to a medical evaluation, potential living donors are required to complete a pretransplant psychosocial evaluation, ideally to be completed by an independent donor advocacy team [43, 44]. Families need to be aware that there are many other nonmedical (e.g. ethical, psychological) factors that need to be assessed, such as the donor’s ability to give informed consent, the donor’s motivation for donating, the relationship between the donor and recipient, emotional factors that may impact donation, and whether the donor has an adequate support system [45]. Further considerations are needed for minors being considered as potential living donors [46, 47], in particular, determining whether there is any coercion involved and whether the minor can appreciate the potential consequences of living donation. The potential negative psychological impact (e.g. feelings of depression, guilt or failure) on potential donors who are not deemed to be a suitable candidate needs to be discussed with families who are considering living organ donation [48].

Families may also assume that all children needing a transplant would view living organ donation in a positive way; however, this area has received little attention within the literature and there are some aspects of living organ donation that can have a potential negative psychological impact on the child recipient. For example, if no one volunteers to donate, or no one is deemed to be a suitable donor, this may result in feelings of sadness or low self-worth in the child. If a suitable donor is identified, some children may simply feel uncomfortable receiving an organ from someone they know or worry about the impact of donation on that individual. If the graft is later lost due to rejection, children who receive their organ from someone they know may feel a strong sense of guilt. All of these potential issues need to be addressed as the child/family heads toward transplant.

Impact of health literacy

When medical information about transplant referral is provided to the patient and family, the concept of health literacy is important to consider. Health literacy is defined as “the degree to which individuals have the capacity to obtain, process, and understand basic health information and services needed to make appropriate health decisions [49]. Research has shown that nearly half of the population in the United States is unable to follow health care recommendations or fully understand informed consent for procedures [50]. This has tremendous implications for families heading toward transplant, a treatment option which requires a very complex evaluation process as well as a very complicated follow-up medical regimen. Little is known about health literacy in pediatric patients/families referred for transplant. However, in a study conducted at an adult kidney transplant center, 41% of patients had limited health literacy [50]. Other studies of adult kidney transplant recipients have found that many patients have gaps in transplant knowledge [51, 52]. A study of adolescent kidney transplant recipients suggested that acquiring more knowledge about the transplant experience helped them to cope; however, individuals varied in the ways they wanted to be educated about the transplant experience [53]. Thus, it is important for medical centers to carefully consider how referral information about transplant is presented and the subject is discussed.

Decision making and informational needs of families

The importance of involving young people in decision making in transplantation and the need to address issues of autonomy, consent and assent have been highlighted [54]. However, the focus in the literature is on decision making at the time of listing and there is no evidence of what consideration is given to these issues at the time of referral for transplant or of the impact on young people of the decision to refer for transplant. The variability in illness chronicity and impact (physical, developmental/cognitive, educational, social and emotional), together with other demographic and family factors, supports the need for an “individualized approach” to engaging pediatric patients and families at the time of referral (or ideally before).

The information needs of parents related to their child’s chronic illness and the importance of effective, individualized information provision have been well documented [55]; however, there is little evidence which identifies the needs of parents at the time when transplantation is being considered. In one of the few studies to address this specifically, Lang and colleagues used qualitative methods to elicit the perceptions of parents whose children had been referred for consideration of lung transplantation for cystic fibrosis [9]. They specifically asked about parents’ experiences of being referred, how they thought the flow of information should be managed and how the process of initially introducing a family to the idea of lung transplantation could be improved. The authors found that parents preferred to know about lung transplantation before their child’s health deteriorated, although they acknowledged that this might be anxiety provoking, and they wanted detailed information about the processes and rationale. In contrast to earlier findings [56] parents wanted the specialist nurse (with whom they tended to have a closer relationship) to begin the process of introducing lung transplantation, rather than the physician, and they only

wanted their child to be involved in discussions when they were old enough to understand and be involved in decision-making. Although this study was limited to patients with end-stage lung disease, such an approach is likely to be appropriate in other disease groups where the child has a long-term chronic illness and transplant is a potential option. Data from adult studies indicates the importance for patients of understanding the reasons for referral, the risks and alternatives [7, 10] and this is likely to also be true for the parents of children being referred for transplantation (and the children themselves, if they are old enough).

Studies of parents of heart transplant candidates [7, 57] have identified the significance of understanding the family's illness narrative and individual and family belief systems and values, including issues of culture and spirituality, irrespective of the illness course that brings the child and family to the point of transplant referral. Data from adult transplant studies indicate that factors such as socioeconomic status, ethnicity, income and insurance status can influence transplant referral and listing [58, 59] and within pediatric transplantation racial and socioeconomic disparities have also been identified. [60, 61]. A survey of nephrologists highlighted that parental level of education and patient history of non-adherence were likely to influence decisions to refer for transplant [62] but it is not clear whether such findings are generalizable to other organ groups nor how such findings translate into clinical practice. Furthermore, there is no evidence of how ethnicity, culture and other sociodemographic factors influence parents' and children's engagement with transplant referral and decision-making processes.

Conclusion

Understanding what transplant referral means for children and families is difficult to clarify due to the fact that this area has received limited attention within the literature. The failure to truly consider the referral phase is illustrated by a description of a sociocultural transplant experience model [63] in which the first phase of the model is described as "assessment and candidacy", yet the evidence from the work with the parents of cystic fibrosis patients in particular clearly indicates that the first phase should be the start of the transplant journey, at the point when transplantation is first being considered.

The failure to address the referral phase as an area of significant importance is understandable as referral often times represents the passing of the baton from one medical team to another. However, certain steps can be taken to enhance the referral process for pediatric patients and families. Based on our review of the literature an individualized approach that takes into consideration patient and family demographic, developmental, and cultural factors as well as disease process and medical experience is of significant importance. Preparing families early on for the potential need for a transplant referral is also preferable. If transplant appears to be a clear possibility, providing patients and parents with the opportunity to explore the reasons for potential referral, life expectancy without transplant, the likely timeline of the transplant evaluation process, and the meaning of the referral in regards to ongoing medical management would be helpful. Referring teams are instrumental in setting the stage for a positive experience for patients and families and helping them understand what to expect in addition to clearly defining their role versus the

role of the transplant team. Further, the involvement of mental health providers in this process such as psychologists, social workers, or psychiatrists working with the referring team could provide additional support and allow patients and parents to explore fears and concerns within a safe environment (see Table 1 for a list of suggestions to enhance the transplant referral process). The involvement of mental health providers can also help the referring team determine whether other barriers to referral exist and help assist families in developing plans to effectively manage the referral process. Such providers could then become an integral part of the support structure for the patient and family during the waiting period for transplantation.

One area that we did not touch upon in our discussion is the relationship between the referral team/center and the transplant program. This is an area of interest as medical providers can also vary significantly in regards to their experience with transplant and their understanding of the transplant process. Hence, outreach by transplant programs to referring teams/centers should be an area of consideration when thinking of the referral process and its impact on pediatric transplant patients and families. Without the right tools to guide prospective patients and families referral centers could be at a disadvantage in setting up a successful experience for their patients. Transplant centers can help referring teams/centers by establishing close working relationships that promote open communication and a cooperative environment in caring for patients throughout the transplant journey.

In regards to research there are many opportunities to further examine the transplant referral process. Referral teams could explore patient and family preferences in regards to the referral process and identify factors that may subsequently affect the patient's and family's ability to successfully undergo evaluation and navigate transplantation. For example, it could be that mental health problems or chaotic family environments which are known to affect post-transplant outcomes also affect the referral and subsequent evaluation for transplantation. Referral centers would then be in the best position to provide support to such families (e.g., recommendation of mental health support; participation in mental health treatment at the referral institution) and if medically feasible perhaps even re-evaluate the timing of the transplant referral. Transplant teams could further explore the needs of patients, families and referral centers in regards to early communication about transplantation and needed information, documentation, and expectations in regards to the process.

The transplant journey is a complicated and potentially unpredictable process that requires a long-term view of patient and family needs. Further exploration into the referral for transplantation will help elucidate the factors that may set a patient and family on the right course for being as successful as possible throughout the transplant journey.

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Figure 1.

Number of Pediatric Transplants as of August 2013

* Based on OPTN data since 1988 to August 16, 2013

Table 1

Suggestions to enhance the transplant referral process

Potential steps to enhance transplant referral
<ul style="list-style-type: none"> • Utilize an individualized approach taking into consideration patient and family factors that may make the conversation regarding potential transplantation or the transplant referral difficult or challenging (e.g., stress level within family, health literacy, previous history of nonadherence to care). • Utilize members of the team who are most familiar, comfortable, and connected to the patient and family being referred to initiate the conversation. • Start the conversation early on. If transplantation is a possible treatment course introduce the idea early to allow the patient and family time to adjust to the possibility. • Assess patient and parent resources and ability to cope with a referral early on. Engage mental health providers in the evaluation of these factors and address concerns and/or barriers. • Discuss treatment course and prognosis including life expectancy without transplantation. • Provide patient and family with information about transplantation in a directed manner that will allow them to take in the information without being overwhelmed. Utilize specific talking points while allowing flexibility for questions generated by the patient and family. • Clarify the role of the referral center versus the transplant center in the ongoing management of the patient's medical condition both prior to and following transplantation. • Provide information about potential transplant centers and suggest avenues to obtain additional information taking care to direct families to trusted sources of information. • Engage in an ongoing open discussion of potential treatment courses and assess the understanding of the patient and family regarding transplant options. • Clarify misconceptions or misunderstandings regarding the transplant evaluation or direct the family to an appropriate source of additional information. • Reassess patient and family understanding during subsequent visits.