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**LUNG TRANSPLANTATION**

*What are the issues?*

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**INTRODUCTION**

Since the 1960s, when the first-ever heart transplant was carried out, organ transplants have become one of the great medical advances of the 20th century. Organ transplants now provide real opportunity for improved quality of life and survival, and cystic fibrosis (CF) is a perfect example of this. There is no effective therapy of end-stage pulmonary disease in CF other than lung transplantation. Lung transplantation has been used as a treatment for end-stage lung disease since the 1980s, and by 1999 almost 1,000 children had received lung or heart/lung transplantations worldwide. The majority have been children with CF.

CF patients tend to have better survival rates after lung transplantation, compared to other indications for this procedure. The survival rate of patients one year after lung transplantation is 75-85%. Five-year survival is estimated to be somewhere between 45-65%. However, this assumes that the patient receives the transplant; unfortunately, 40% of patients will die whilst on the waiting list.

Children with CF, especially females, have a higher two-year mortality rate than their adult or male counterparts. Young female patients are even highlighted in the referral guidelines because they have a poorer prognosis and need early referral. This means that a lot of the transplants occurring in CF patients are in children, which introduces many more issues into the scenario, both medical and ethical. This article aims to look at several pertinent ethical issues surrounding the decision for lung transplantation in children with CF.
MAKING THE DECISION – CURRENT LITERATURE

Being faced with the choice of having a lung transplant is a massive emotional rollercoaster. There are so many factors to consider, risks and benefits, quality of life issues, probable outcomes and so much more. How does a ten-year-old child cope with this decision? Do the parents or doctors decide for them? Does the child make the decision? What happens if the child decides that they did not want to go on the transplant list? How does one cope with the waiting game of being on the list? It is an ethical and emotional minefield. It is a situation no one would wish to be in, and a decision no one would make without difficulty, especially for their own child.

So when deciding whether to say ‘yes’ to going on the transplant list there are a number of issues to consider. If one agrees to proceed with the transplant, and is then placed on the active transplant list, they then have to cope with the stress of actually being on the list. They will always be waiting for that call with a bag packed, they cannot plan holidays, they always need to be contactable, and they have to be careful about what analgesia to take. This process of waiting does not just impact upon the person with CF, it affects their whole family, including their siblings, and will have an unknown impact on the family’s quality of life. Being on the list does not guarantee that one receives new lungs; the patient may die whilst waiting for that call. In addition, we must not forget that the procedure itself is major surgery and thus comes with its own risks, and so the patient may be one of the few who does not survive the operation. In this case, life is cut shorter than it may otherwise have been, with or without the transplant. A successful lung transplant does not guarantee longterm survival; due to complications, the patient may not survive much longer than they would have done without. Post-transplant, some people are glad for the life they have been given, and make the most of every extra moment they have. Others are cautious about contact with others, aware of their relative fragility, worried about infection or rejection. Some patients suffer chronic rejection (obliterative bronchiolitis), so their quality of life is affected. However, we must not forget the success stories, and the reason why people want the transplant in the first place. With a transplant, the patient may have a completely transformed life, they will no longer have CF lungs, and they will hopefully be able to do things that they have not previously been able to do. They may survive for many more years to come. For some people post-transplant, the time spent with family and friends and doing things they had never experienced far outweighed any risks. If they make it through the first three years post-lung transplantation then over 90% of children have no limitation to their activity.

Some people decide not to go on the list for transplantation, and choose instead palliative care. This choice immediately eliminates the risk of the procedure, the risk of complications and the risk of organ rejection. They may know approximately how long they will have left, and they will be able to put their affairs in order, possibly plan their own funeral, and say their goodbyes. They may not have the expectant waiting that a lot of people experience on the list, they will be able to make the absolute most of the last days they have left. For some people, this is the right decision and they are happy with the lifetime they have been given and the time they have left.

The patient and family cannot make a fully-informed decision regarding transplantation unless they are given all the information in detail. Legally and ethically this is a necessity. Children with CF are recommended for transplantation based on three factors: life expectancy of two years or less, poor quality of life, and no contraindications for transplantation. With only two years of life expectancy, the child and their family are at an extremely vulnerable stage. The glimpse of possibly restoring a meaningful quality of life or extending a child’s life may inhibit the family’s realistic conception of the whole transplantation process. Discussing lung transplantation at this highly emotional time may introduce doubt about influencing capacity, voluntariness, and comprehension. Before a decision is made, the patient and family must be completely informed of the entire transplant process, including patient selection, indications, donor criteria, surgical technique, longterm follow-up and outcomes. It must also be emphasised that lung transplantation is not a cure for CF but trades one set of problems for another. ‘the ability to breathe is traded for the complications of life-long immunosuppression’. The psychological and emotional state of both the patient and the family can influence the degree to which they embrace or fear transplantation, and the child will be greatly influenced by the doctor and parents’ interactions.

The recently published General Medical Council document, 0-18 years: guidance for all doctors, discusses age and consent to treatment and is recommended to all readers. The decision to label someone competent is based upon their ability to understand the relevant information, their ability to believe the relevant information and their ability to weigh up the information and make a decision.

Should this be adhered to in the case of lung transplantations in children? If one leaves the decision up to the parents then this may be going against the wishes that the child expresses. To survive and thrive after lung transplantation the patient must be motivated to get better and to comply with all the necessary medication; this may not be so easy if the child did not want the treatment in the first place. Leaving the decision to the parents also puts a lot of pressure on them. If the child does not survive the operation, if there are complications or if their quality of life is not improved, they may blame themselves, and live with resentment from their child. However, if a relatively young child is to be left with the final word how can we be sure that they are truly competent to make this decision; do they really understand all that is involved?

It may not be useful to consider a strict dichotomy between competence and incompetence, but rather a sliding scale of competency may be more helpful. By adolescence, compliance with therapy depends almost solely on the patient, rather than the parents, and involving these patients in decisions regarding their care is necessary to maximise compliance. However, some still believe that more onerous decisions, such as those involved in transplantation, should remain with the parents until the child reaches the legal age of consent.

It is thought that children are limited in their comprehension of disease until about the age of nine years, and even up to the age of 15 children may not identify preferences of their own that have not been influenced by authority figures. Thus, whether a child’s decision is ever
truly voluntary may always be questionable. However, children with CF may fall into a slightly different category. These children have had plenty of past experience with medical therapies and procedures. Previous experiences influence a child’s ability to participate more fully in decision making, and these children may therefore have an advantage in this process.\(^{(9)}\)

The transplant team at Great Ormond Street Children’s Hospital let the child have the final word, respecting their autonomy. They believe that this is the only way that the child will be motivated to get better and that it gives the child the best chance of survival.\(^{(9)}\) There is a well-known connection between emotional wellbeing and compliance with medication and treatment.\(^{(10)}\) The outcome will always be better if the patient has a positive outlook on life with a strong emphasis on sought-after goals.\(^{(9)}\)

**CONCLUSIONS**

Lung transplantation is not a cure for CF lung disease. At best, it is a procedure which can provide a child some extra years of life, with improved lung function allowing them to undertake previously impossible activities. At worst, it can shorten life, or provide false hope as children await a transplant that will never occur.\(^{(9)}\)

Lung transplantation in children with CF introduces many ethical issues. Capacity and competence are brought into play when deciding who can give consent for such an important procedure. Voluntariness is also a vital element of consent, ensuring that it is a legitimate decision, and that there is no coercion or other forces acting on the involved parties, influencing the decision. Autonomy is very important, especially when considering children. To respect a child’s autonomy in this situation would be to let them make the decision or have the final word. Best interests are also relevant, and can be very subjective. What the child sees as in their best interests might not be what the parents would consider to be in the best interests of their child.

Beneficence also comes into play. The beneficent act may appear to be to give the child the lung transplant (or rather to put them on the list giving them the opportunity to have the lung transplant), as this would seem to be the action adhering to the definition of beneficence – to do good. However, it does not come without risks, and this is why patients are only referred when these risks are outweighed by the benefits. The patient must be ill enough to need the transplant, but well enough to survive the surgery.

All these ethical principles need to be considered when faced with a family in this situation, as well as remembering the legal position. If a family is pro-transplant, but the child is not, then a decision for transplantation is going against the child’s autonomy; the child may not see the decision as being in their best interests, but the family may see it as the beneficent option. Issues of consent do not just apply to the child; with such an emotionally charged issue, the parents’ voluntariness may be difficult to assess. Everyone is different, and every family is different. It is the child’s life that is under discussion, and surely they should have a huge part to play in any decision that is made regarding it.

**REFERENCES**


9. Will I still love my mum? Channel 4 Television documentary. 3 August 2005


