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TRANSITIONING THE YOUNG ADULT WITH AN INHERITED METABOLIC DISEASE

Inherited Metabolic Diseases (IMD) are a rare group of disorders of metabolism leading to an imbalance of chemicals. This can negatively impact organ systems which may have irreversible effects to development.¹

As some of these disorders are identified on newborn screening and treatment is optimised early, this group are surviving beyond their adolescent years and require planning for their long-term health requirements.

Despite many official reports from the National Service Framework for Children,² Royal College of Nursing³ and Royal College of Paediatrics and Child Health⁴ highlighting the need for a smoother transition of care, not many departments have the correct models to assist in this process. Transition is best described as the purposeful, planned movement of adolescents with chronic medical conditions from child-centred to adult-orientated health care.⁵ The process should acknowledge not only the medical needs of the young adult, but also their psychological, social, educational and overall wellbeing.

The ultimate goal is to empower the young adult to make informed decisions in relation to their health outcomes, and support them in reaching their full potential in society.⁶

CHALLENGES

There are many factors that impede the transition process, especially as the IMD patient requires management of complex issues, such as modified diets, varying regimes of medicines, physical, mental and/or learning disability affecting daily living, access to emergency care, fertility and inheritance of their genetic condition. It is a multidisciplinary team approach often requiring community input.

A lack of engagement from the young adult and their family can make transitioning very challenging. This may be caused by their lack of understanding for the rationale behind this process. One American source reported that less than half of parents had discussed the health needs of their child with their health provider.⁷ There is a danger that the young adult may become lost to follow-up if there is no clear guidance regarding the transfer of their care.

Within my centre, transition is a four-staged process with introduction to the adult IMD team at the third stage. Even after the fourth appointment where both teams review the young adult, patients remain under the care of the paediatric team until they have been seen solely in the adult setting. This is to prevent misunderstanding of who the care provider is between appointments, where the patient is 'in limbo'. Miscommunication, conflict and contradictory advice must be avoided during this period so that the young adult does not feel that neither the paediatric nor the adult team are responsible for their care.⁸

The bond between the paediatric team and the young adult and their family will have been established from early on. The paediatric physician may have been the person who diagnosed the rare IMD condition, with the paediatric multidisciplinary team offering support to the family during the distressing initial stages of diagnosis. The team will have gone through a journey with this family; from diagnosis to schooling

Table 1: Appropriate age for transition⁷

Age (years)	Adolescents with special needs (%)	Adolescents without special needs (%)
<12	3	2
12-14	6	6
15-17	25	26
18-20	62	65
Don't know	4	2

Does not total to 100% due to rounding.

achievements, making it difficult to break the bond.⁷ Conversely, if this bond has not been established and oppositional behaviour occurs with non-compliance from the parent and patient, abrupt transition may appear tempting, so the young adult can then establish a fresh start with the adult team.⁸ However, it may result in the young adult feeling rejected by their paediatric carers and may prevent trust being established with the new care provider, the adult team.

In order to see the paediatric physician, who may still be the most suitable caregivers, young adults may not wish to attend a children’s hospital, or to find themselves waiting in a room with toys. This can lead to non attendance⁶ due to embarrassment and disengagement as the young adult may feel that his/her needs are not being adequately met.

What is the correct age for transition? Should it be when the physician feels that they are ready? Table 1 shows data from 628 respondents from the American Academy of Paediatrics to a questionnaire regarding the age at which paediatricians think transition planning should begin.⁷

It has been reported that some services use a cut off age range of 15 to 20 years, and some use events such as school leaving as a trigger for transition.⁸ A systematic review identified two studies transferring patients at aged 18 and eight studies ranging between ages 16 to 20.⁹ The authors suggested using tools to measure self-management and advocacy skills for preparation of transition instead of age⁹ to avoid inconsistency. It is clear that the timing of transition is paramount and variable; however, it should not be one event, but a series of processes.

The geographical location of the adult service can be a deterrent to families who are accustomed to the paediatric location.⁹ Transition should not

be avoided or delayed, even with both services in close proximity. Complex IMD is managed by other teams such as cardiology, liver, renal, neurology and/or dental, which may not be at the specialist IMD centre.

In my experience, it can be very stressful for the family when transferring to more than one adult specialty at one time. However, a disparity of care and access occurs when the young adult remains under local paediatric gastroenterology, for example, having accomplished a smooth transfer into adult IMD services.

Another barrier to transition may be the lack of specialist adult physicians, which particularly affects IMD as this is an evolving service. Many young adults are seen by the paediatric physician, clinical nurse and dietitian as there is no adult service to refer them onto. Some IMD dietitians have a split role managing both paediatric and adult patients. The young adult will then be familiar with the existing team and there may be reluctance to transition if the staff and location remain the same. Nonetheless, adult-related topics should be explored with the young adult and patient involvement should be encouraged.

PATIENT EXPERIENCES

Focus groups have been used to assess the young adult’s experience of transition. The themes recognised after transcribing their interviews from various literature of those with long-term health conditions were categorised into the following headings:

- Expectations of the transition¹⁰
- Meeting the new team¹⁰
- Life decision support¹⁰
- Support from others¹⁰
- Timing¹⁰
- Physician mistrust¹¹



Table 2: Strategies for successful transitions

Focus	Intervention	Rationale	Strategies
Young adult	Education on their condition	Self-efficacy	One-to-one teaching ⁹
		Increase confidence	Visual resources
		Achieve responsibility ¹²	Peer support groups
			Internet based teaching
	Informed decision-making	Patient involvement ^{3,4}	Motivational interviewing
			Agree goals on transition documents and timeline to achieve ³
Personnel	Named transition link ^{3,6,9,13}	Continuity of care	One point of contact ^{3,9,13}
		Structured transition process	Attend all transition clinics
		Prepares young adult	Offer a visit to the adult setting
	Coherent administrative support	Prevent young adult being lost between appointments	Checklist before transfer ^{6,8}
			Set transition clinics in the diary for the year
	Training by Royal Colleges ⁴	Increase awareness and skills	Mandatory within the curriculum for allied health, nurse specialist and trainees
Service	Joint paediatric and adult clinic	Gradual introduction to adult services ⁹	Adult team to attend paediatric clinics
		Improve information sharing	Letters to be addressed to the young adult
	Young adult clinic	Diminishes feeling lost amongst general clinics	Set up young adult clinic on a different day to other clinics
	Out of hours contact	Approachable team offering support at times convenient to the young adult	Provide telephone contacts for emergency care
	Enhance follow-up	Prevent consecutive non-attendance	Contact the young adult to enquire rationale for non-attendance
	Include community services	Continuity of care	Open channels of communication
			Engagement with the General Practitioner

- Difficulty with employers¹¹
- Stress in personal relationships^{11,12}
- Moving from a familiar to an unknown ward culture¹²
- Being prepared for transfer¹²
- Achieving responsibility¹²

One quote from a young adult which implies lack of experience and interest of the physician was the following:

“Dr X is my primary doctor but he doesn’t really know anything about the disease, so I have to tell him what to do.”

Emotions during transition were also examined and coded in qualitative literature describing feelings of abandonment¹¹ and a feeling of letting go without knowing what the future entails.¹²

Positive experiences around inclusion of decision-making and being treated like an adult include quotes such as:¹²

“At the paediatric centre they’d talk to the parents and say, ‘you must make sure your child takes medication’. At the adult centre, they tell you the benefits of it, they tell you what happens if you don’t take it and leave it in your hands, so they give you a lot of control...they do talk to you like you’re an adult; it is your decision.”

“The most important thing to me when transferring to the adult clinic was that all the decisions were made with me involved and I was able to talk about what was going to happen when I saw the doctors, which I never had a chance to do in the children’s clinic.”

It has been suggested that these positive experiences should be shared with paediatric patients preparing for transition¹¹ to manage expectations.

RECOMMENDATIONS

Generic and disease specific transition models or programs have been described.^{2,8} Within my centre, a specific IMD transition model was created which can be applied for use to other chronic conditions. The Welsh IMD service identified geographical gaps in transition.¹³

Whichever model is adopted, the principles for successful transitions should include these valuable suggestions summarised in Table 2.

Physicians in the adult services should avoid making drastic treatment changes and extensive investigations⁸ at the first appointment to prevent feelings of unease and to manage expectations of the young adult and their family. The physician is required to build a rapport confidently whilst engaging the young adult.

Health professionals ought to be ready to invest the necessary time and support to prepare young adults to take responsibility for their own health. Allowing sufficient time for consultations, active listening by the multidisciplinary team and anticipation of young adults’ involvement may enhance their experience of themselves as valuable contributors.

CONCLUSION

Long-term outcomes of IMD patients receiving medical and dietary treatment is undefined. Therefore, transition from paediatric to adult services should be a gradual and planned process with engagement from the young adult and their family. Special consideration is required when transitioning the young adult with complex needs. Transition champions have been proposed as part of a gold standard within the skilled multidisciplinary team.^{8,9} The transition process should encompass a systemic outlook covering not only health, but also vocational and psychosocial aspects.^{9,12}

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