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## THE PRINCIPLES OF THE TRANSITION PROCESS FROM PAEDIATRIC TO ADULT SERVICES IN INBORN ERRORS OF METABOLISM – OWN EXPERIENCE

### ZASADY PROCESU PRZEJŚCIA PACJENTA ZE SZPITALA DZIECIĘCEGO POD OPIEKĘ LEKARZA DOROSŁYCH – DOŚWIADCZENIA WŁASNE

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#### Abstract

*The transition from the paediatric to the adult service is a complex process that requires early planning to prepare young adults and their families for the transfer to the adult hospital. The article outlines the main principles of the successful transition period. Importantly, a young adult becomes an independent individual, interacts with medical professionals independently, and makes the appropriate choices. Both healthcare professionals and parents should facilitate the process of building new relationships between the adolescent and the adult metabolic teams.*

**Key works:** transition, Inborn Errors Metabolism, self- direction

#### Streszczenie

*Proces przejścia pacjenta z zakładów opieki zdrowotnej dla dzieci do szpitala dla osób dorosłych jest procesem złożonym, który wymaga planowania tak by młodzi pacjenci i ich rodziny były przygotowane na czekające je zmiany. W niniejszej pracy opisano główne zasady prawidłowej organizacji okresu przejściowego. Przede wszystkim, osoba staje się niezależną, samodzielnie nawiązuje kontakt z pracownikami Służby Zdrowia, i powinna być zdolna dokonywać właściwego wyboru. Zarówno lekarze, pielęgniarki jak i rodzice powinni umożliwić proces budowania nowej relacji między osobą młodocianą a zespołem opiekującym się dorosłymi z chorobami metabolicznymi.*

**Słowa kluczowe:** proces przejścia, choroby rzadkie, samodzielnie ukierunkowany

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#### THE TRANSITION PROCESS

Due to advances in medicine and developments in research over many recent years, significant numbers of young adults with metabolic conditions are able to survive into adulthood. Many rare metabolic conditions were only described in paediatric textbooks, whereas

nowadays they continue into adult life. *‘Transition is the purposeful, planned movement of adolescents and young adults with chronic physical and medical conditions from child-centred to adult- oriented healthcare systems’ [1].*

The transition from the paediatric to the adult service is a complex process that requires early planning to prepare young adults and their families for the transfer to the adult

care. This article outlines the golden principles of the successful transition process based on personal experience from our Adult Inherited Metabolic Disorders Centre. The age of consent, legal requirements to be an adult and the ages at which young patients become adults depend on the legal and cultural measures in different countries. In this review the example relates to the legal and other requirements as practised in England where the age of consent is 16, most paediatric services end at the age of 18, and the Mental Capacity Act of 2005 [2] determines how patients with limited capacity should be treated.

### Principle 1 – Consent

The young adult is encouraged to learn all about their condition and that they may alert health professionals when medical help is required. The young adult will require skills and education to manage their illnesses, often independent of their parents. They will be required to give consent and make decisions about their own healthcare, such as giving their permission before they receive any type of medical treatment or undertake any research projects. If an adolescent lacks the capacity to give consent, a capacity assessment will take place to assess if an individual is able to understand the information, retain this information, then use or weigh up this information to make a decision and communicate the decision [3].

The mental capacity of patients with rare metabolic conditions has an impact on the amount of input that they have in the transition process [4]. The fact that a patient has limited mental capacity should not stop healthcare professionals from taking into account their wishes and choices wherever appropriate [4].

Some individuals with learning difficulties (their IQ is less than 70) are not able to make simple/complex decisions due to impaired adaptive or social functioning, which refers to a reduced ability to cope independently [3].

### Principle 2 – Passport

The hospital passport is a patient's document outlining the pathway plan and that is initiated at the beginning of the transition (fig. 1). Both the patient and the transition team contribute to the document to transfer medical notes to the adult hospital in preparation for transfer care [3]. When the young patient becomes an independent individual, the hospital passport facilitates the monitoring of the continuity of care and Paediatrics patient's compliance with specific therapies such as enzyme-replacement-therapy infusions, betaine, ammonia scavengers and aminoacids exchanges. Any significant medical events and data should be captured in the document and ensure appropriate the handover between paediatric and adult metabolic teams. Examples of events documented include the dates of the last insertion of venous access devices and the due dates for their replacement, skeletal and cervical imaging to ensure the appropriate management in the new unit. Additionally, all patients who turn 16 years of age and have the capacity need to renew their consent to continue having their data included in the registries. To avoid any confusion, it is crucial that the transition documents are completed before a patient with a known Inborn Error of Metabolism is referred to adult services.

For example, a referral of a Mucopolysaccharidosis patient who turned 18 years of age to an adult Ear and Throat Specialist cannot precede their transition to the adult metabolic team in the same hospital.

### Principle 3 – Relationships

The integral part of the transition process is the need for the patient to become an independent individual, to interact with medical professionals independently, and to make appropriate choices [4].

Initially when the child is young, the relationship is between the parents and the medical team, with the child a passive observer [4]. The transition process that can take two to four years is the start of a new relationship that is built up between the young patient and the medical team, with the parents becoming passive observers. Finally, the young patient becomes completely independent from the parents and manages their own healthcare. This process can be very difficult, especially when the parents find it difficult to let go and are not prepared for the changing relationships.

Adolescent patients should be encouraged to be part of the process as much as possible. They should be presented with options to make their own choices, even if they are different from the choices their parents might make [4]. It is important that a young patient has some time alone with the transition- team members, although many of them need a family member or a friend to attend at the beginning or end of the consultation.

### Principle 4 – Adherence

Adherence means the extent to which patients follow medical treatment and advice. It is estimated that around 30-50% of medicines prescribed for long-term illnesses are not taken as directed. It is detrimental to patients and healthcare systems, with estimated double the risk of hospitalisation and financial costs. The main reasons for non-adherence are lack of information/lack of perceived benefit and/or lapse of memory [5]. The unintentional non-adherence that accounts for 30% of cases can be explained by forgetting or poor communication between patient and healthcare professionals. The intentional non-adherence (70% of cases) results from patients' unwillingness to adhere to treatment regimes [5].

There is a high risk of young adults not adhering to treatment or failing to adapt to the adult system. This can result in preventable additional complications with increased healthcare utilisation e.g. admissions to High-Dependency Units and a decreased quality of life [6]. Adherence is lower among adolescents than adults, particularly as they approach independence. Support from their parents and school plays a vital part in their self-image and indirectly in the adherence to therapy. Achieving therapy goals requires that patients have a simple but correct understanding of their condition and are aware of what goals will be achieved and how that will improve their quality of life.

Complex, rare, metabolic conditions require a multidisciplinary team to assess their severity in specialist centres in terms of special diets, medication/supplements and the timing and frequency of clinical monitoring.



Young adults feel overwhelmed by the intensity of care and changes around the transition to adult services, particularly in that they might have never felt unwell and they have never felt the necessity to adhere to, for example, sick-day rules. Additionally, they often present their concerns regarding peer compliance [5]. Peer groups have been shown to be the best motivators in comparison to healthcare professionals. Additional measures are to see if drug-dosing intervals can be changed as once-daily dosing is more acceptable than twice-daily dosing and in young adults with Tyrosinaemia moving to once-daily dosing can improve adherence, even if the evidence is limited as once-daily dosing is better than missing repeat dosages [7]. Many young people are using mobile phones or other devices that can be set up to create reminders and forming closed peer-support groups on social media is allowed in some countries [8, 9].

### Principle 5 – Self-Directed Management

The transition to adult health services requires significant self-direction for which many adolescents are not adequately prepared or willing to take on [6]. Self-direction requires some readiness to make their own decisions regarding health and full awareness of all the normal adult healthcare issues such as weight management, the safe use of alcohol, osteoporosis, fertility and cardiovascular health. It is important for a young patient to have an insight into the consequences of the poor control of their chronic condition, such as female patients with Phenylketonuria before they plan pregnancies. As often happens, pregnancies are unplanned in the majority of cases and future mothers are not frequently compliant with treatment during the peri-conception time.

Condition-specific and generic self-management skills are very important. From the motivational point of view, it is better if young adults themselves come up with solutions, although they still have support and encouragement from the clinic staff. There is evidence that Self-Directed Management (SDM) improves the long-term outcomes of patients during the transition process. This hangs on some simple principles where young people are encouraged to talk about what is important to them, with the motto *“It is OK to talk about anything”*. This includes body image, relationships and other issues which might be more important to young people. Specifically designed prompt sheets can be made to cover topics that will be age, culturally and country specific. One of the methods that is also encouraged is the use of 3 questions system:

1. What are my options?
2. What are the pros and cons of each option for me?
3. How do I get support to help me make a decision that is right for me (10)

Self-management is the management of themselves and their learning enterprises. Choices and freedoms are matched by self-control and responsibilities. Patients learn to express self-control by searching for, and making a commitment to, core personal interests and aspirations. It requires confidence, courage and determination to energise the effort involved.

Self-determination is a concept reflecting the belief that all individuals have the right to direct their own lives. Young adults who have self-determination skills have a stronger chance of being successful in making the transition to adulthood, including employment and independence [11, 12]. Self-determination helps young adults with disabilities achieve positive adult outcomes. A person's actions are self-determined if the person acts autonomously, regulates his or her own behaviour, initiates and responds to events in a manner indicating psychological empowerment, and behaves in a manner that is self-fulfilling [12]. A self-determined person is one who sets goals, makes decisions, sees options, solves problems, speaks up for him/herself, understands what supports are needed for success, and knows how to evaluate outcomes [13].

This example supports the above. An 18-year-old male MPS IVA patient attended the transition clinic at the children's hospital twice, and on both occasions, his family were both heavily involved in the consultation and he spoke very little. However, during the consultation the adult team emphasised the importance of taking control of his own healthcare and to work towards SDM for the future. On the day of his second appointment, he attended the transition clinic in the morning with his dad and again spoke very little with simple 'yes' and 'no' answers. In the afternoon of the same day he came across to the adult services and was seen by the adult team who directed all questions to the patient and encouraged him to respond. He was much more confident and answered the questions fully. It was suggested by his consultant that he needed to have a 5-day cardiac monitor and due to the distance he would need to travel that he might be better doing this locally, as he would need to collect it and return it after 5 days. The patient suggested that he would prefer to have it done at the expert centre, as they would have more experience to interpret the tests. He chose to travel down on public transport with his dad to return the tape by himself a week later. He wanted to know how he could get to the hospital by himself and we discussed the options with him, i.e. train, bus, taxi etc. Then he mentioned that he was looking to move out of his parents' house and move into an adapted house. Parents were taken by surprise but were very supportive as they realised that although he has not been expressing his views to them he had already researched and collected information in his own time. We also discussed the fact that he was applying for a provisional driving licence and wanted to learn how to drive. We plan to encourage him to be seen without his parents during his next appointment. The change in him was remarkable and we hope that this continues as time goes on. This taught us the lesson that although young adults do not always interact in the transition process the moment you create the ideal circumstance they can suddenly take advantage of it and the time spent in transition is well worth it for the majority of patients.

### Principle 6 – Autonomy

Meeting the adult team also gives the patient a good opportunity to discuss issues that they do not want to

mention to their parents, e.g. sex, contraception, planning pregnancy, or the safe use of alcohol and drugs. Practical topics such as taking a gap year, how to manage the enzyme replacement therapy during their placement in Australia, or how to adjust doses of steroids/amino acid exchanges after a long flight can also be raised.

The adult team should always attempt to have one of the multidisciplinary team speak to the young person alone. Even patients with learning disabilities need to be assessed and for part of the consultation they can be alone, as long as more than two professionals are present.

One of ways to achieve this is to get a professional to talk to a parent whilst another talks to the young person. Alternatively, we ask parents to leave while we examine the patient.

Importantly, we set the tone on entry by addressing the young person or by asking them if they want a parent to leave.

Young adults require constant support during the transition so that they can interact as a competent adult and fully take over their own care, e.g. book appointments with their local GP.

### The Principles of Effective Transitions\*

1. A written policy, including patient-held medical records
2. The personal involvement of the young person in the process
3. A preparation period and an education programme
4. A coordinated transfer process with a named coordinator
5. Transfer on a planned date when ready in both services, if possible
6. An interested and capable adult clinical service
7. Good communication throughout the process
8. The involvement of patient organisations or groups to promote self advocacy
9. Administrative support
10. Primary health care and social-care involvement

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### Author's contributions/Wkład Autorów

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\*Adapted from Hendriksz CJ. (2013) [14].