

The challenge of transition in chronic respiratory diseases

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Transition – what does it all mean?

Transition has been defined as the “purposeful, planned movement of adolescents with chronic medical conditions from child-centred to adult-oriented health care” [1]. Paediatricians see chronic health conditions in 10% of their patients, 90% of whom will survive to adulthood [2,3]. An increasing awareness of the need for transition services for youth with chronic health conditions has resulted from advances in medical technology and treatments that have increased the lifespan and quality of life of these youth [4].

Why should we care?

Chronic conditions in childhood are prevalent

◀ 16-31% have at least 1 chronic illness

◀ 2-7% have serious disability

◀ Over 7 million children have asthma

◀ >90% of children born today with severe chronic medical conditions will live to adulthood

◀ ~ 750,000 adolescents with chronic conditions are expected to transfer from pediatric to adult care [5].

Therefore, respiratory diseases represent an important percentage of the paediatric chronic medical conditions. In addition, the incidence of allergic disease and asthma has increased over the last 20 years and this rise may continue. Asthma is the most common cause of school absence, and cystic fibrosis (CF) the most common lethal inherited condition [6].

Adolescents with chronic respiratory illness

Respiratory symptoms such as cough, sputum expectoration and breathlessness are physically but also socially embarrassing. Patients may feel isolated and ashamed if they cannot keep up in sports or attend trips and take time off. These “visible” symptoms may result in bullying from peers and overprotection by parents and teachers [6]

Other conditions are obstructive sleep

apnea secondary to childhood obesity and bronchopulmonary dysplasia which in the future each will need specific transition programmes. Several conditions, such as primary ciliary dyskinesia, non-CF bronchiectasis and sleep disordered breathing are currently underdiagnosed, leading to preventable morbidity. Children under the age of 17 years are identified as receiving long term ventilation (LTV) and other children receive some kind of respiratory support at home.

Fundamental differences exist between paediatric and adult chronic care

Paediatric care is often multidisciplinary, prescriptive, and family focused. It requires parental direction and consent.

The chronic care model involves a multidisciplinary team that provide coordinated care based on planned visits with *follow up* telephone and electronic contact, and a focus on monitoring disease markers, delivering preventive services, and coaching children and family members in disease self-management.

Adult care tends to be patient focused, and it encourages autonomy in making decisions about treatment and life choices. Professionals in adult care are familiar with the difficulties associated with sex, pregnancy, work, and raising a family in the context of chronic ill health.[7]

Respiratory conditions in childhood are increasing in prevalence and need good transition units. As an example, for children with asthma, there is often a lack of clear diagnosis and most are seen in primary care. It is well known that adolescents with asthma do not get referred to the adults services and disappear into a “void”[6]

Poorly planned transition from young people's to adult-oriented health services can be associated with increased risk of non adherence to treatment and loss of follow-up, which can have serious consequences. There are measurable

adverse consequences in terms of morbidity and mortality as well as in social and educational outcomes. When children and young people who use paediatric services are moving to access adult services (for example, during transition for those with long term conditions), these should be organised so that all those involved in the care, treatment and support cooperate with the planning and provision to ensure that the services provided continue to be appropriate to the age and needs of the person who uses services[6,7]

Rare diseases - still rare disease, but no longer orphans

Historically, rare diseases were also called orphan diseases. They were orphans because no one was developing therapies for them. There was no chance that a company would be able to recover its investment if it spent time and money developing a treatment for such a small population.

Some of these diseases are no longer 'orphans' in this regard, because the bio-pharmaceutical industry is now 'adopting' them slowly, but steadily [8]. In this case, the paradigm of cystic fibrosis (CF) care has changed as effective therapies extend the lives of patients well into adulthood.

Advances in cystic fibrosis (CF) treatment have changed CF from a disease lethal in early

childhood to a chronic illness with a median survival of 40 years [9]. Emerging small molecule therapies hold promise to further extend survival [10].

Patients with cystic fibrosis describe their experience of living with the disease from childhood to adulthood. Among the many challenges they describe is the "rocky road" of transition from paediatric to adult health care. Sometimes they say that they would have given anything to attend a transition clinic when they were 16 years old, instead of going straight to an adult clinic at another hospital. [7]

Transition to adult care for any child with a chronic life limiting illness should not consist of just transfer to a doctor who treats adults. It should be a clinical and psychosocial process [7].

Challenges to transition exist on various levels:

- ◀ Patient
- ◀ Family
- ◀ Provider
- ◀ System

Patient Challenges

- ◀ Neurocognitive development has not fully matured in most adolescents
 - ◀ Patients with chronic diseases have an abrupt expectation at "age of majority" to undertake their own care
 - ◀ Pediatric patients often do not have the skills to self-manage their own care
 - ◀ Only 40% of CSHCN report receiving any guidance [McManus et al, Pediatrics 2013]
- Aging out of child programs determines loss of insurance, increased use of emergency services and loss of primary care, having significant unmet medical needs.

Parent Challenges

- ◀ Parents are ill-prepared to navigate the adult health care system and adult programs
- ◀ Parents lose legal rights of their children at age of majority
- ◀ As parents age, they may not be able to care for their child

Provider Challenges

There are some differences in provider roles

- ◀ Pediatrics much more family centered (or patient excluded)
- ◀ Adult Medicine much more patient centered (or family excluded)
- ◀ Differences in subspecialist vs primary care roles

Very often there is a poor or non-existent communication between pediatric and adult care providers.

What do patients and their families want?

Adolescents want: continuity of care and early introduction to the adult team; team knowledge of disease and individual medical history; honesty and confidentiality; a named staff member and telephone access; to be seen privately; clinic hours that are less disruptive; to sit away from other age groups or those with severe infections. Initially they find shorter consultation times difficult, but notice other benefits, such as being treated with maturity and respect. Parents remain more concerned than children about transition [11,12,13]

Transfer of Information

Transfer of information and multidisciplinary team reports must be accurate and complete. An example from CF is annual review data, which

includes sputum microbiology, spirometry, oxygen saturations and body mass index. If patients see a new thin

folder in place of their previously thick one, they may wonder whether the new team knows their complete medical history. Joint transition clinics can avoid this [6].

Transition Timing

Discussions about transition can begin at any time; however, it is best to begin this process by the time a child is 12 years old. Discussions about transition are best held during a checkup or routine chronic illness visit.

Transition to an adult-oriented clinic typically should occur within the 18- to 21-year age range. There are cases in which transition can occur earlier if the adult clinic accepts new patients at younger ages and cases in which transition can occur later if transition goals are not yet reached[14].

Preparation for adult roles takes time

Most pediatricians focus on the last visit, but acute issues arise making this event difficult.

Transition planning is a purposeful process, screening patients and families to make sure everything is in place before the transfer event.

Not all aspects of care have to change at the same time: primary care transfer, subspecialty transfer, school changes (going off to college, work, living).

Who Is Involved in the Transition?

Successful transition involves the participation of the whole medical team, including physicians, nurses, and other care coordinators. The medical team works with the family and the child to create goals and a timeline for transition. It is important to identify each team member's role in the transition to help the child to begin to take on a more independent role in his or her health care.

Parents' role is to be part of the process but to start to step back from making every decision so that the child gets increasing experience and independence making health care decisions[14].

Conclusion

Successful self-management of asthma and food allergies is still challenging. Persistent sleep difficulties may be associated with a number of health, economic and emotional difficulties and rise risk of suicide in sleep-deprived teenagers [15]

Those with childhood chronic illness utilise healthcare for most or all of their lives. This group

of adolescents and young adults require a system that serves them appropriately, in their transition from paediatric to adult patient, as they continue their lifelong fight against disease. This is a global phenomenon which requires set gold standards. Transition services have been developed for children with other chronic conditions, such as diabetes, renal disease, and complex congenital heart disease in addition to transplant recipients. The principles are similar, although local resources and the underlying condition determine the details of care.

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