Transitioning the Pediatric Pulmonary Hypertension Patient

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Advances in disease awareness, earlier diagnosis, and additional therapeutic options for treatment of pediatric pulmonary hypertension (PH) in the last 2 decades have dramatically improved survival, leading to the first generation of pediatric patients surviving to adulthood.[1-4] Pediatric care is generally a family-centered approach, whereas adult care is more patient-centered. When patients become young adults they must move from dependency on parental involvement and oversight to independence and individual accountability. It is difficult for young adult patients to navigate this transition successfully without coordinated support from their family, pediatric, and adult care providers.

Recent analyses of pediatric PH within REVEAL and in the Netherlands Registry demonstrate the 2 primary subgroups as idiopathic pulmonary arterial hypertension (IPAH) and PH associated with congenital heart disease. IPAH is seen less often in children, while PH associated with congenital heart disease is seen more often than in adults.[4]

The proportionately higher incidence of young adults with PH associated with congenital heart disease that will be receiving care through adult PH programs in the future will require incorporating adult congenital heart disease (ACHD)–trained physicians into the adult PH care team. The ACHD community has formally addressed the need to create more structured programs with a recent publication of a best practice statement for managing transition to adulthood for adolescents with congenital heart disease.[5] The adolescent medical community has recognized issues around transition for more than 2 decades, including a position paper published by the Society of Adolescent Medicine in 1993.[6] Transition practices can be further modeled from large pediatric chronic disease populations that have already piloted and implemented these processes, such as cystic fibrosis and sickle cell disease.[7-9]

Traditionally, adolescent patients have transferred to adult programs through a "drift-away" model: an incomplete, vague transition from the pediatric team instead of a clear and comprehensive "handoff" to the adult care team.[10] The "drift-away" model of transfer has been an unsuccessful transition, leaving young adults struggling to manage their disease and treatment well, and frequently resulting in being lost to follow-up.

Over the past 5 years, the center at Children's Hospital Colorado has experienced increasing numbers of adolescent PH patients achieving college acceptance, entering the work force, beginning to live independently, and therefore moving away from their nuclear family and support system. Parallel to this, young adults are required to transition from a dependent role where their parents led interactions with health care providers, coordinated medication refills and dosing, and, for those on invasive therapies, often mixed and changed the infusions daily. In addition to the usual challenges of entering college or the work force, these young adults suddenly have to take ownership of their disease process, become independent in medication administration, identify changes in clinical symptoms, maintain medication compliance, and learn to access the medical providers and arrange clinic follow-up appointments. These challenges have led to the development of a transition program that will assist providers, young adults, and their families in making a structured yet individualized, comprehensive, and successful transition to an adult program.

Barriers to successful implementation of a transition program include insufficient staffing, lack of identified staff members responsible for transitions, financial challenges, institutional acceptance, and resistance from the adolescents and their parents in transferring to an adult center. Commitment to partnership and open dialogue between pediatric and adult programs is vital for smooth and individualized transitions of care.[5,10,11]

Key aspects of the transition process include: timing; patient, family, and provider readiness; identification of adult PH care team; successful completion of transition curriculum; and transfer of care (Figure 1). The American Academy of Pediatrics, American Academy of Family Physicians, and the American College of Physicians have recommended that the transition process start as early as 12 years of age, with the physical or absolute transfer of care between 18 and 21 years of age depending on developmental readiness.[10] Even before the actual education or curriculum portion of the process begins, the concept should be discussed with the patient and family. This decreases stress of the unknown, as many pediatric patients have built a good rapport with their pediatrician and pediatric care team.

After developmental readiness is assessed and the patient enters the transition process, the curriculum focuses on basic understanding of diagnosis and sequentially builds to medication management,

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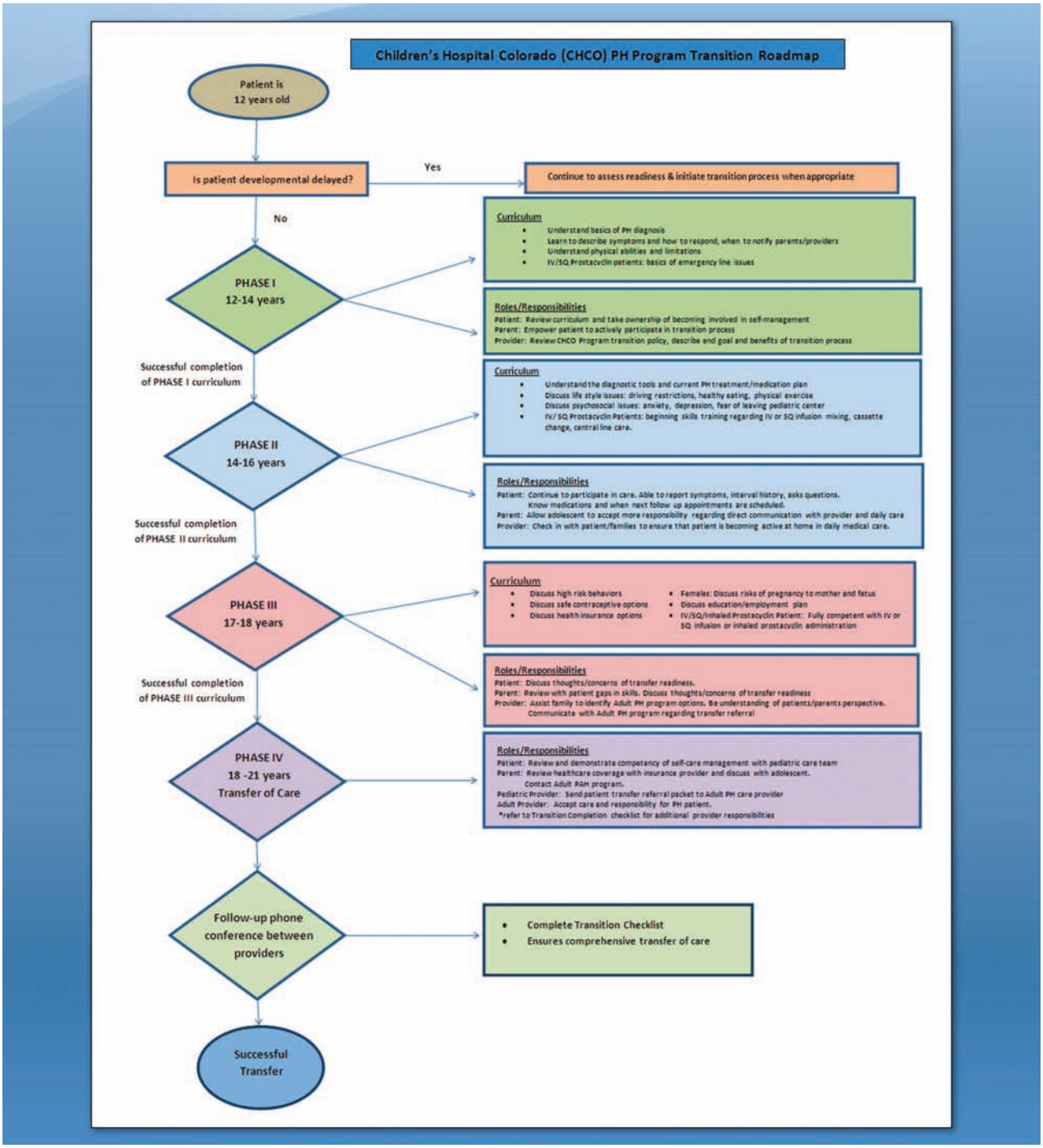

Figure 1: Transition Road Map.

infusion therapy, and direct interaction with the medical care team. Parents play a critical role in the early stages of the transition process, as they must be committed to the process and willing to start relinquishing control, empowering the patient to work toward independence in health care management. The expected age of transfer, roles, and responsibilities during transition should be identified, discussed, and provided in writing.[10]

It is important to recognize and discuss patient and parent needs and perspectives during the transition process, understanding that it is complex and potentially emotional for them. Families of chronically ill children have made significant changes and accommodations in their lives to care for their children. The pediatric care team has been a constant, is familiar, and is known. To be successful the young adult has to be the driver throughout the process, nourishing self-management skills and growing autonomy.[5]

In conclusion, transition planning should be a standard policy in pediatric PH programs to maximize patients' abil-

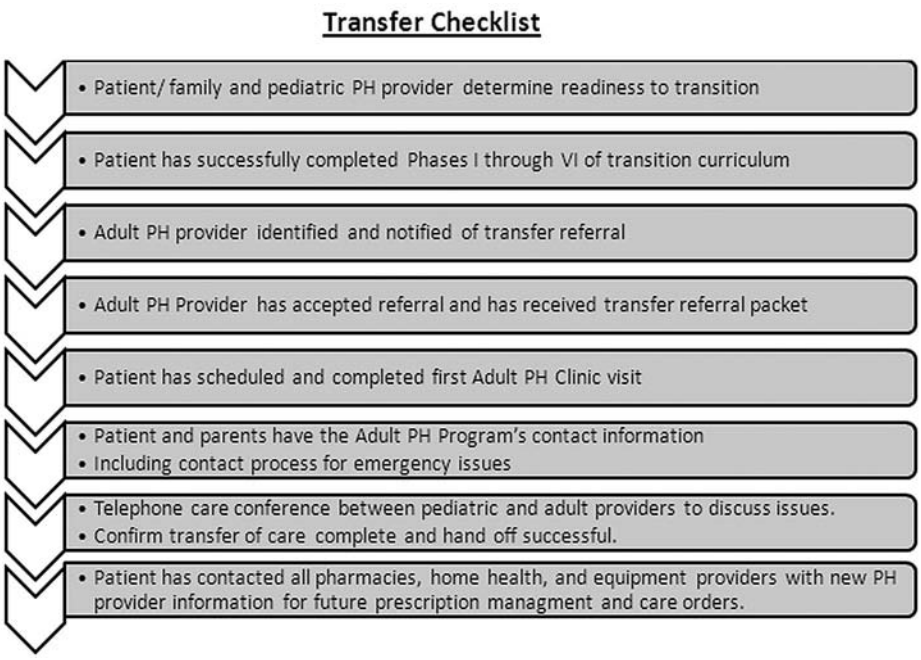

Figure 2: Transition Checklist.

ity to successfully care for their chronic health needs as they enter adulthood. Recognizing the barriers and forming partnerships with adult PH programs will result in increased numbers of young adults successfully completing the transition process with transfer of care to the adult center.

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