

Child and Family Centered Care for Pediatric Patients Diagnosed with Pulmonary Arterial Hypertension

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Tremendous advances have occurred in the treatment of childhood pulmonary arterial hypertension (PAH) over the past decade. However, PAH remains a challenging, chronic, and potentially life-threatening disease, with an estimated 5-year pediatric mortality rate as high as 28% currently.^{1,2} The prevalence of PAH in children is estimated at ≥ 3.7 cases per million.³ PAH is characterized by a gradual increase in pulmonary artery pressure that ultimately can lead to right heart failure and death. Comparable histopathological changes within the pulmonary vasculature are noted in adults and children. However, compared with adults with similar severity of disease, cardiac output is better preserved in children.^{1,3} As a result, in children, ordinary activities of daily living may not cause symptoms. The most commonly reported symptoms of dyspnea and fatigue may only be present with over-exertion and are more commonly associated with syncope.¹ Thus, when a child presents with minimal symptoms associated with daily activities, it is often traumatic for the child and family to face the sudden reality of the diagnosis of PAH.

In addition to the sudden shock of the PAH diagnosis, the family and child face difficult and life-changing decisions regarding the medical management of the disease. The management of PAH may have a detrimental impact on the child's and family's normal daily routine due to multiple hospitalizations, frequent clinic visits, and therapies. Also, there are often other complicated issues to be considered such as role changes within the family to allow for care and close monitoring of the

child, the economic impact of the cost of medical care and lost wages by caregivers, and emergency preparedness at home and school.⁴ In some cases, the stress may even lead to divorce. Some PAH treatments are very complex and may involve continuous parenteral infusions or multiple daily dosing of oral and/or inhaled therapies.¹ The child and family may suddenly experience significant stress as they come to terms with the reality of the severity of the diagnosis and its complicating impact on their lives.

Research addressing the psychological impact of PAH in adult patients is just beginning to emerge and has demonstrated an increased incidence of depression and anxiety.^{5,6} In addition, a recent study that evaluated the impact of adult PAH on family caregivers found an increase in depressive symptoms among the caregivers. The study noted a correlation between the increase of the depressive symptoms and a lack of sufficient emotional support and information to meet the demands of caregiving. However, interestingly, caregivers also perceived their responsibilities as positive and rewarding and a source of self-esteem.⁷ No studies have been conducted on the impact of PAH on pediatric patients or their families.

Healthcare professionals involved in pediatric PAH recognize the stressors and challenges faced by the child and family as they learn to integrate the child's illness and medical management into their daily lives. The literature on pediatric chronic illnesses also recognizes the long-term challenges faced by the child and family facing such an illness.⁸ For example, there are multiple ongoing processes of adjustment and integration for the child, siblings, parents, extended family, peers,

school personnel, and the community. Poor adaptation to chronic illness has been associated with psychological problems such as poor self-esteem, adjustment disorder, depression, as well as family dysfunction.⁹

As the family is the child's primary source of strength, support, and stability, the American Academy of Pediatrics (AAP) supports the healthcare model of family-centered care as a means to maximize the well-being of the child and family. While there is no single definition for family-centered care, at its core is the philosophy that healthcare professionals and the family are partners working together to meet the needs of the child. This family-centered approach includes the principles of dignity, respect, information sharing, participation, and collaboration (Box).¹⁰

The role and involvement of the child within the discussions and treatment decisions will vary depending on the child's stage of cognitive development. Following diagnosis of PAH, initial discussions may be best with the parent(s) alone followed by a developmentally-appropriate conversation with the child and eventually with the siblings. Actively involving a child in his or her own healthcare is also supported by the AAP and has been shown to improve parent and child satisfaction.¹¹ Such child involvement increases the child's knowledge of the prescribed therapy, and improves overall functional status.^{11,12} As adherence with PAH therapies is critically important, allowing the child the opportunity to be a part of the process and included in decisions appropriate to his or her cognitive development can provide the child with some sense of control and ownership.

Knowledge of the ways that children understand disease and the concepts of life and death is essential for healthcare professionals and families in order to

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Principles of Family-Centered Care¹⁰

- 1) Treat patients and families with dignity and respect
- 2) Communicate unbiased information
- 3) Support and facilitate choice for the child and family about approaches to care
- 4) Child and family participate in experiences that enhance control and independence and build on family strengths
- 5) Collaborate in the delivery of care, policy, and program development

communicate more effectively with and meet the needs of the child. Various theories have been presented over the years, many of which are consistent with the Piagetian model of cognitive development. In the Piagetian model, there are 4 major developmental stages: sensorimotor (0-2 years), preoperational (2-7 years), concrete operational (7-12 years), formal operational (12+ years).^{13,14} A child in the first 2 stages of the Piagetian model understands illness in a prelogical manner, meaning that external objects or people can “magically” cause illness. They may also see illness as punishment for wrongdoing. In the next stage, concrete operational, the child may still view the cause of illness as being external. However, the child now recognizes that the illness is located within the body. At the same time, the child experiences confusion about internal organs and their function. In the formal operational stage, the child understands that illness may be caused by external or internal events, and recognizes the process of cause and effect. The child sees illness as a step-by-step process that culminates in a malfunctioning internal organ. In this stage, a child can also appreciate the influence of psychological factors such as stress or happiness as having an influence on illness in general.¹⁵

In this Piagetian developmental model, a child’s understanding of illness and death proceeds in a step-wise fashion, closely tied to his or her cognitive developmental maturation. More recent theories have allowed for mobility between the suggested developmental stages, as a child may not be firmly located at one stage or adhere to only one concept of the illness.¹⁴ Understanding these cognitive

stages of development and assessing the child’s view of the illness and therapy can assist healthcare clinicians and families to guide the child gently through new information and ideas. During follow-up, it is important to re-assess the child’s understanding in light of his or her cognitive development and address any increasingly complex questions and concerns. A strong relationship between the child and the healthcare team can provide a safe environment for the child to express his or her concerns. Research shows that children with cancer can exhibit anxiety, depression, low self-esteem, adjustment disorder, and fears related to dying.^{16,17} The healthcare team must be alert to these and other issues and work with the child and family to address any problems.

The well-being of the family is the foundation of family-centered care.¹¹ Families face many new challenges and stressors as the result of the child’s diagnosis of PAH. From the broader literature, it has been noted that demands placed on a parent as caregiver can result in depression, anxiety, sleeplessness, and poor health.¹⁸ Siblings are also susceptible to increased anxiety, insecurity, and loneliness, especially during the first 6 months following diagnosis.¹⁹ Research examining the impact of a chronically ill child on the marital relationship has demonstrated negative effects such as role strain²⁰ and a decline in sexual relations.²¹ However, positive effects on the marital relationship have also been noted including improved communication, better conflict resolution, and an increase in trust.²¹ Although research on divorce is very limited, there does not appear to be a disproportionately high divorce rate among parents of chronically ill children.^{22,23} Family structure

variables associated with higher psychosocial risk include single-parenthood, adolescent parenthood, and having 4 or more children.²⁴

The healthcare team can be a valuable resource for families. Family-centered care can improve family stability, empower parents, and enhance self-esteem as parents learn to juggle multiple new tasks and roles. Healthcare professionals can help reduce stressors by providing instruction and guidance, offering access to social and professional support systems, and encouraging positive personal coping mechanisms and cultural values already in place.²⁵ Working toward “normalization” of life is key, including assisting the child to return to school.

Implementing the concepts of family-centered care in pediatric PAH requires a dedicated team of healthcare professionals, including physicians, nurse practitioners, nurses, respiratory therapists, social workers, and chaplains. The work can be exciting and rewarding, but can also be challenging and emotionally draining. Connecting families with a pediatric palliative care team can be another way of helping to meet the needs of the child with severe PAH and his or her family. Palliative care often has been misunderstood as end-of-life care. However, there has been a recent shift in this paradigm demonstrating that palliation can now be combined with curative therapies from the time of diagnosis of a life-threatening disease.²⁶ The reality is that the clinical course for PAH can be unpredictable and suddenly impacted by unexpected events leading to death. Connecting families with the palliative care teams in the early stages of treatment provides an opportunity for relationships to strengthen and allows additional services for the child and family. The palliative care team composed of physicians, nurses, social workers, spiritual advisors, and others can provide such services as art therapy for the child and siblings; assistance with counseling, finances, or travel; blood draws and symptom assessment; and connection with spiritual services.²⁷

In conclusion, family-centered care, developmental psychology, and the broader literature on the impact of chronic and

life-threatening illnesses on children and families can provide helpful frameworks for healthcare professionals to identify and meet the psychosocial and spiritual needs of children with PAH and their families. However, research is still needed to identify and clarify the specific psychological and social impact of PAH on children and families in order to meet their needs more effectively.

Disclosures: Ms Doran is currently employed as a Medical Science Liaison with United Therapeutics and was previously a member of the pulmonary hypertension team at Children's Hospital of Colorado. This article does not represent the opinion of United Therapeutics.

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