Hospitalizations of Children With Pulmonary Hypertension: Implications for Improving Care

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Pulmonary hypertension (PH) complicates the clinical course of children with diverse cardiac, pulmonary, hematologic, and systemic diseases and remains a challenging problem. Despite the growing availability of PH-specific drug therapies, the outcomes for children with severe PH generally remain poor, and the lack of effective treatments further contributes to significant morbidities and diminishes the quality of life.^{1,2} Overall, pediatric PH has been understudied, and little is known about its natural history, longitudinal course, optimal diagnostic approaches, or treatment. In addition, we currently have limited understanding of disease-specific mechanisms in pediatric PH; there is a lack of organized, multidisciplinary care programs; and few markers or end points are available for assessing clinical course and response to interventions in children.³ Importantly, few data address the overall impact of pediatric PH on the health care system, including rates of hospitalization and health care resource utilization.

In this issue of *Pediatrics*, Maxwell et al⁴ provide important new information that examines national trends in hospitalizations and resource utilization as obtained from a national database. The authors report significant rates of hospitalization that have been increasing over the past decade and are associated with rising hospital costs. This study highlights the substantial and growing health care burden of pediatric PH and suggests

possible improvements in morbidity and mortality associated with inpatient PH care. However, the latter findings are especially difficult to fully assess: it is likely that more patients with diverse forms of PH or milder PH severity may have been increasingly included over this time period. This study enriches our understanding of the scope of the problem of pediatric PH and provides data for reflection of ongoing issues.

As noted by the authors,⁴ a potential limitation of the study is the questionable reliability of diagnostic billing codes from the Kids' Inpatient Database to accurately reflect the true frequency of PH-related hospitalizations and related costs. However, this important report will likely stimulate and be followed by more in-depth work. Such studies will include novel approaches that more thoroughly analyze electronic health records and other sources of data to expand on how the nature and costs of childhood PH are defined. More importantly, however, is the exciting potential of novel informatics strategies to further explore the nature of disease in diverse settings, as well as a more comprehensive view of resource use and related questions. Such informatics strategies have the potential to study childhood PH in greater detail, including its related diagnostics, drugs, outcomes, costs, and other issues.

It is likely that data from this report⁴ represent "the tip of the iceberg" regarding the true impact of PH on

hospitalizations, resource utilization, costs, and clinical outcomes. The actual health care costs and resource needs of pediatric PH, as with other chronic diseases, are limited by a lack of data linking inpatient with outpatient care, in which frequent clinic appointments and diagnostic evaluations are essential for improving outcomes well beyond inpatient care alone. The impact of pediatric PH may also be underestimated in these data because the diagnosis of PH and its evaluation are complex, and the role of PH may not be recognized in some clinical settings by health care providers. Despite growing recognition and awareness of PH in many settings, such as prematurity, Down syndrome, obstructive sleep apnea, sickle cell disease, oncologic disorders, chronic lung diseases, liver disease, and others, PH is often a “hidden” contributor to morbidities and outcomes. In addition, even mild PH may reflect pulmonary vascular disease that contributes to greater susceptibility regarding the need for hospitalizations for viral respiratory infections, exercise intolerance, and recurrent respiratory problems, especially in children born prematurely and children with Down syndrome.

Further studies are also necessary to define the relative costs and benefits

of specific diagnostic or interventional approaches for children with PH. Experts in PH are needed to work with hospital leadership to develop guidelines for cost-sparing policies, as currently exemplified by concerns regarding excessive charges for potentially useful medications (eg, inhaled nitric oxide) for which charges continue to escalate unabated.

Finally, of marked interest is the observation that many children with PH were managed at sites other than children’s hospitals and that these findings suggest a clear lack of centralized care from experienced teams of PH providers.⁴ As previously noted, developing strong interdisciplinary PH care programs at experienced centers and collaborations among these centers will foster high-quality research, standardize diagnostic and treatment strategies, and enhance the development and application of care guidelines.³ As best demonstrated by the marked success of the Cystic Fibrosis Foundation in improving outcomes for subjects with this disease, the need for developing clinical and research networks by using similar approaches may translate into substantial gains for families and children with PH. More research is needed to best define an optimal PH center for

children, but it is likely that this goal will require creation of interdisciplinary teams of cardiologists, pulmonologists, intensivists, neonatologists, and other subspecialists, along with approaches that enhance collaboration between centers.

ABBREVIATION

PH: pulmonary hypertension

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