Study evaluates quality of life and the burden of care with Prader-Willi syndrome

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Researchers from the School of Medicine at Trinity have evaluated the impact of the rare genetic disorder, Prader-Willi syndrome on the quality of life of the affected child, and their family.

In the study of caregivers of children ranging from less than 1 year to 18 years, dietetic and paediatric researchers investigated the impact of weight gain on health and wellbeing in patients with Prader-Willi syndrome. The study was carried out alongside a research team from Children's Health Ireland (CHI) at Tallaght University Hospital.

Prader-Willi Syndrome is a complex genetic condition which causes neurological, hormonal, behavioural, developmental, and cognitive problems. The condition was first described in 1956 and is caused by the absence of certain genes on chromosome 15. It can lead to excessive eating and life-threatening obesity, as well as obsessive/compulsive behaviours. More than 100 cases of this syndrome have been diagnosed in Ireland.

Professor of Paediatrics at Trinity College, Edna Roche said, "We found that Prader-Willi syndrome impacts significantly on quality of life in both the affected child and the family. In addition, we found that quality of life in the patient and the entire family is negatively impacted by increased weight."

Results showed that parents of teenage children with Prader-Willi syndrome reported an increased burden of care made worse by psychological difficulties, and when the child's routine or social activities were disrupted and restricted. Findings also showed parents perceived increased weight and age of their child to have a significant negative impact on their child's psychosocial health and social functioning.

The study highlighted the need for appropriate respite services for affected families.

Professor Roche explained, "Provision of appropriate respite care for children and young people with Prader-Willi Syndrome along with

increased support for their families will not only enhance the healthcare outcomes of affected patients, it will improve their quality of life and that of their families. Importantly it will also enable families to continue to provide care within the home environment."

The study was financially supported by the National Children's Hospital Foundation and was a collaboration between researchers at the Department of Nutrition & Dietetics and the Department of Paediatric Growth, Diabetes and Endocrinology in CHI Tallaght, as well as the Department of Paediatrics at the School of Medicine, Trinity College Dublin.

Commenting on the research, Marguerite Hughes from the International Prader-Willi Organisation said, "This study is a welcome addition to the growing volume of research highlighting the impact of Prader-Willi syndrome on quality of life and the need for support, not alone to children with the syndrome, but also their parents and entire families. The impact on families points to a clear need for social care services (such as respite) to be provided to families at an early stage. The complex needs of children with Prader-Willi syndrome also suggest that social care services must be tailored specifically to the needs of people with the syndrome to be effective."

More information: Christina Meade et al. Prader-Willi syndrome in children: Quality of life and caregiver burden, *Acta Paediatrica* (2020). DOI: 10.1111/apa.15738

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