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New genetic cause of a childhood kidney cancer discovered

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Genetic mutations in a gene called REST have been shown to cause Wilms tumour, a rare kidney cancer that occurs in children.

Wilms tumour affects about 1 in 10,000 children, but fortunately is curable in about 90% of them.

A study led by researchers at The Institute of Cancer Research, London, identified mutations in the REST gene in 16 children with Wilms tumour.

Nine of the children were the only members of the family to develop Wilms tumour, but in four families more than one child had developed the cancer.

It was the clustering of cases of this rare cancer that alerted the researchers that a hereditary genetic cause was likely. They estimate that REST mutations cause about 10% of familial Wilms tumour.

The research is published today (Monday) in the journal *Nature Genetics*, and is part of the Factors Associated with Childhood Tumours (FACT) study, which is uncovering genetic causes of childhood cancers and has participants from more than 5,000 families. The FACT study is funded by the Wellcome Trust and the Rosetrees Trust, and the REST project involved international collaborators including from Baylor College of Medicine in Texas, US.

REST is a well-studied gene because of its critical role in embryo development, but this research brings a new, previously unrecognised, role of REST to the field of human genetics.

The mutations found in children with Wilms tumour appear to compromise the normal functioning of REST in regulating the development of the embryo.

Study leader Professor Nazneen Rahman, Head of Genetics and Epidemiology at The Institute of Cancer Research, London, and Head of Cancer Genetics at The Royal Marsden NHS Foundation Trust, said:

"We hope our findings will stimulate research into why and how these REST mutations, which all cluster in a particular part of the gene, cause cancer.

"Our findings are also of immediate value to families, who now have an explanation for why their child got cancer. Moreover, we can now do a simple blood test to see which children in the family are at risk of cancer and may benefit from cancer screening, and which have not inherited the mutation and so are not at increased risk of cancer. This kind of information is really valuable for the families of children with cancer."

More information: Mutations in the transcriptional repressor REST predispose to Wilms tumor, DOI: 10.1038/ng.3440

Provided by Institute of Cancer Research

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