Research studies at Newcastle University 2014

Researchers at Newcastle University have been carrying out two studies on patients with histiocytic diseases.

Adults with histiocytic disorders in Northeast England

The first study, on adult disease, began in 2012. Adults may present to many different clinical specialities depending on the site of their disease and they do not have an established diagnostic or treatment pathway. There is no national or international agreement on the best treatment and no open clinical trials or national registry.

There are currently 4 adult clinics in the UK, specifically for patients with LCH and related disorders - a long-established one at the Hammersmith Hospital in London, one at the Royal Victoria Infirmary in Newcastle, one at the Leicester Royal Infirmary in Leicester and one at the Royal Hallamshire Hospital in Sheffield. Although patients with LCH are seen at the clinic in Newcastle, little is known about adults with other forms of the disease. Efforts have been made to increase the awareness of the histiocytoses and encourage collaboration among clinicians but the scale of the problem in the UK is unknown.

Researchers are trying to establish how many adults there are with these diseases in the Northeast region and also identify to which specialty patients were referred, and the treatment and services they received for their disease or any long-term disabilities. It is hoped that the results of the study will encourage co-operation and planning for patients both regionally and nationally so that patients benefit from all the available expertise. It is also hoped that the methods used will be expanded to a national level to establish a registry of patients.

Cases have been identified from all 8 regional hospitals in the Northeast and further information on presentation, treatment and outcome is being collected from treating clinicians. The most common disorders were juvenile xanthogranuloma and Langerhans cell histiocytosis with a small number of cases of Rosai-Dorfman disease, Erdheim-Chester disease and haemophagocytic lymphohistiocytosis. In addition, cases of fibrous histiocytoma and reticulohistiocytosis were identified. The study, which is funded by the JGW Patterson Foundation in Newcastle upon Tyne, is expected to end in May 2015.

Langerhans cell histiocytosis (LCH) in children with congenital anomalies

The second study, which was completed last year, investigated the presence of congenital anomalies in children with histiocytic disorders. Congenital anomalies have been associated with childhood cancers and although they have been noted in patients with Langerhans cell histiocytosis (LCH), only one small hospital-based study over 20 years ago had previously examined their frequency. An association between the two may suggest a common origin, e.g., a genetic mutation or predisposition, or common environmental origin. Alternatively, children with severe congenital anomalies may be at a higher risk of developing histiocytosis, e.g., as a result of physiological changes, lifestyle changes, or possibly treatment they may have received.

This study investigated this possible link by identifying children with both conditions using two well-established, population-based registries in the North of England - the Northern

Region Young Person's Malignant Disease Registry and the Northern Congenital Abnormality Survey.

The results of the study were presented at the annual international Histiocyte Society meeting in 2013 and a publication is currently under review. The study was funded by Newlife Foundation for diasabled children.

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