INCIDENCE, SUB-SPECIALISATION AND THE PROVISION OF DEFINITIVE SURGERY FOR HIRSCHSPRUNG'S DISEASE IN THE UK AND IRELAND: RESULTS OF A PROSPECTIVE, NATIONAL COHORT STUDY

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Aims: 1) Determine the incidence of Hirschsprung's Disease (HD) in the UK and Ireland; and 2) Calculate the impact of current sub-specialisation on Consultant case volume in the delivery of definitive surgery for HD.

Methods: Between October 2010 and September 2011, pre-determined reporting clinicians at each paediatric surgical centre in the UK and Ireland returned monthly reporting cards, prospectively identifying any live-born infant presenting before 6 months of age, with histologically proven HD. Incidence with binomial exact 95% confidence interval was calculated using total live births as the denominator. Total number of Consultant Paediatric Surgeons and the proportion offering definitive HD surgery was established through questionnaire with follow-up email/telephone call. Potential number of cases per surgeon was calculated for each centre. The Mann-Whitney test was used to compare groups.

Results: During the 12 months, there were 169 confirmed cases of HD in the UK and Ireland in neonates and infants <6 months of age, giving an estimated incidence of 1.9 per 10,000 live births (95%CI 1.6-2.2). Emergency general paediatric and neonatal surgery is provided by 160 consultants across 28 regional centres, with 110 consultants performing definitive HD surgery. Consultants working in 17 centres with designated HD surgeons potentially perform a median of 2 cases/surgeon/year (IQR 1.1-2.8), compared to 1 case/surgeon/year in centres without designated HD surgeons (IQR 0.6-2.1, p=0.03). Nationally, 56 surgeons at 12 centres (51%) will perform definitive surgery less than or equal to once per year, and 34 surgeons at 11 centres (31%) will perform 2 or more cases per year.

Conclusion: This is the first time the incidence of HD in the UK and Ireland has been calculated prospectively using data collected from the entire population. There are regional variations in sub-specialisation and delivery of definitive surgery. Despite sub-specialisation, only 31% of surgeons involved in HD will perform 2 or more definitive procedures per year.