

## British Association of Paediatric Surgeons Congenital Anomalies Surveillance System (BAPS-CASS)



The national system to study the surgical management of a range of neonatal conditions

## Newsletter 13 July 2010

# **New Study – Hirschsprung's Disease**

Hirschsprung's Disease (HD) is a common cause of intestinal obstruction in the newborn period, characterized by an absence of ganglia in the enteric nervous system of the distal bowel (aganglionosis). The estimated incidence of HD is 1 in 5000 live births<sup>1,2</sup>, with 90% of cases diagnosed in the newborn period.<sup>3</sup> However, there is limited information about the incidence and mode of presentation of HD in the UK.<sup>4,5</sup>

This study will be the first national, population-based study with prospective follow-up of a birth cohort of infants with HD. It will provide accurate data regarding the incidence and associations of HD in the UK; the current surgical management of HD; and complications and specialist service utilization of these patients in the first year of life. These data will provide a benchmark against which paediatric surgeons can assess their own operative results, and hopefully lead to improved outcomes for patients with Hirschsprung's Disease.

The study, funded by Newlife charity will be led by Gregor Walker and Tim Bradnock from Glasgow Royal Hospital for Sick Children, and will commence in October 2010.



- 1. Spouge D, Baird PA. Teratology 1985; 32: 171-177.
- 2. Suita S, Taguchi T, Ieiri S, et al. J Pediatr Surg 2005; 40: 197-201.
- 3. Singh SJ, Croaker GD, Manglick P, et al. Pediatr Surg Int 2003; 19: 247-250.
- 4. Bodian M, Carter C. Ann Hum Genet 1963; 26: 261-277.
- Orr JD, Scobie WG. Br Med J (Clin Res Ed) 1983; 287: 1671.

#### **Case Report Summary to 07/06/10**

Condition	Cases Reported	Forms returned	Confirmed Cases	Unconfirmed Cases	Duplicate Cases
CDH	171	106 (62%)	103 (97%)	1 (1%)	2 (2%)
Oesophageal Atresia	184	158 (86%)	153 (97%)	3 (2%)	2 (1%)

### **Star hospitals**

Many thanks to the following units who have returned all the data for the oesophageal atresia study:

Addenbrooke's Hospital Bristol Royal Hospital For Children Chelsea and Westminster Healthcare Trust Edinburgh Royal Hospital for Sick Children Glasgow Royal Hospital for Sick Children Hull Royal Infirmary King's College Hospital Norfolk & Norwich University Hospital Queen Charlotte's & Chelsea Hospital Queen's Medical Centre Roval Aberdeen Children's Hospital Royal Liverpool Children's Hospital Sheffield Children's Hospital Southampton General University Hospital Lewisham University Hospital of Wales

And to the following who have also returned all data for the CDH study to date:

Bristol Royal Hospital For Children
Edinburgh Royal Hospital for Sick Children
Hull Royal Infirmary
Norfolk & Norwich University Hospital
Queen's Medical Centre
Royal Aberdeen Children's Hospital
Royal Alexandra Children's Hospital
Royal Liverpool Children's Hospital
Southampton General

Addenbrooke's Hospital

Thank you!

## Meet the team – BAPS-CASS at the BAPS International Congress

Various members of the BAPS-CASS team will be at the Congress in Aberdeen to discuss and present completed and forthcoming studies. Sean Marven will be manning a 'trade' stand where you can catch him to discuss gastroschisis and all things BAPS-CASS, Kant Shah with study lead **David Burge** will present the first results from the oesophageal atresia study, Gregor Walker and Tim Bradnock will excite your interest in the new Hirschsprung's disease study, Paul Losty will be on the hunt for CDH data and Paul Johnson will also be available to answer questions.



Marian Knight/Sean Marven (Clinical Coordinators): 01865 289727

Carole Harris (Administrator): 01865 289714

email: BAPS-CASS@npeu.ox.ac.uk web: www.npeu.ox.ac.uk/BAPS-CASS