Pseudotumor cerebri Syndrome (PTCS) in childhood: current management and one-year follow-up of a national prospective population-based cohort study

Yim-Yee Matthews[1], Fiona Dean[2], Ming Lim[3], Karen Mclachlan[4], Alan Rigby[5], Guirish A. Solanki[6], Colin Kennedy[7], Cathy White[8], William P Whitehouse[9, 10]

[1]Paediatrics, Wrexham Maelor Hospital, Wrexham; [2]Ophthalmology, University Hospital Coventry & Warwickshire NHS Trust, Coventry; [3]Children's Neurosciences, Evelina Children's Hospital, London; [4]Paediatrics, University Hospital Coventry & Warwickshire NHS Trust, Coventry; [5]Hull York Medical School, University of Hull, Hull; [6]Paediatric Neurosurgery, Birmingham Children's Hospital, Birmingham; [7]Clinical Neurosciences, University of Southampton, Southampton; [8]Child Health, Morriston Hospital, Swansea; [9]School of Medicine, University of Nottingham, [10]Paediatric Neurology, Nottingham Children's Hospital, Nottingham, UK

Objective

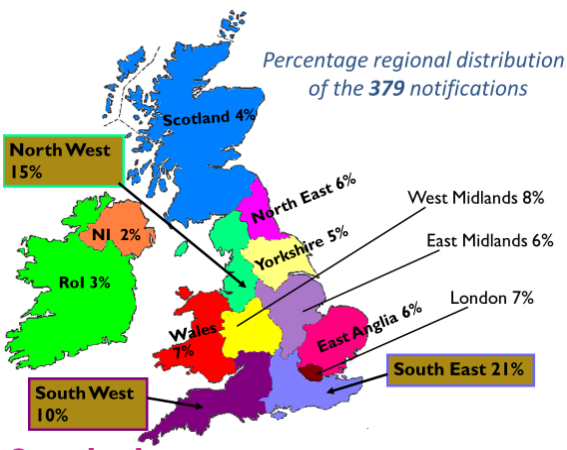
To ascertain current management of children and young people aged 1-16 years with pseudotumour cerebri (PTCS) in the United Kingdom and Republic of Ireland, and their outcomes 1-year after diagnosis.

Methods

A national prospective population-based cohort study was conducted over 25 months 2007-09. Newly diagnosed PTCS cases, ascertained using classical diagnostic criteria, were notified to the British Paediatric Surveillance Unit.

Data on incidence, clinical profile, and risk factors has been published: Matthews et al. *Arch Dis Child* 2016: doi: 10.1136/archdischild-2016-312238.

Data was also collected on treatments used and 1-year outcomes post-diagnosis.



Conclusion

Significant nationwide variation in management was observed reflecting the lack of high-quality clinical trials in PTCS. Despite having the lowest incidence, children aged 1-6 years with PTCS had the highest rate of neurosurgical intervention, and the greatest risk of visual impairment.

Clinical trials are urgently needed to inform the best treatment approaches in PTCS in childhood.

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https://www.rcpch.ac.uk/about-us/our-team/british-paediatric-surveillance-unit-team

and all our colleagues who notified likely cases and then provided more detailed information.

Results

185 newly diagnosed PTCS cases aged 1-16 years were identified, from 379 notifications.

Treatments given

Summary of Interventions	No.	%
Self resolved with no intervention	9	**5**
Stop Minocycline, Prednisolone or Growth Hormone	4	**2**
Medical treatments	155	**84**
Acetazolamide (9 or 5% experienced adverse effects)	151	82
Furosemide or Coamilofruse	30	16
Prednisolone or Dexamethasone or Methylprednisolone	13	7
Topiramate	10	5
Therapeutic Lumbar Puncture(s) only	10	**5**
1 to 2 LPs	150	81
3 or more LPs (max >50)	35	**19**
Neurosurgery & Neuroradiology	24	**13**
Shunting procedures: LPS (16), VPS (8), VAS (2)	26	
Optic nerve fenestrations (1)	1	
Cranial vault expansion (2), subtemporal decompression (1)	3	
Venous sinus stent (1)	1	
Had more than one surgical procedure	5	**3**

24 who had neurosurgical procedures included 13/124 (10%) females and 11/61 (18%) males. Younger children were more likely to undergo surgery: based on age-category, 8/17 (47%), 8/58 (14%), and 8/110 (7%) who had neurosurgery were diagnosed between 1-6, 7-11 and 12-16 years respectively.

Weight loss was reported as an additional intervention in only 6/185 (3%) of cases.

1-year follow-up data was submitted on 163/185 (88%), including on 113/163 (69%) females.

Outcomes 1-year post diagnosis

Clinical outcomes	At diagnosis		1-year post diagnosis	
	No.	%	No.	%
Headache	161 / 185	87	44 / 158	**28**
Papilloedema	165 / 185 6 unknown	89	37 / 158 4 unknown	**23**
Visual defects	61 / 185	33	16 / 158	**10**
No visual data	5 / 185	3	5 / 158	3
Too Young or Learning Difficulty	2	1	0	0
Don't know	3	2	5	3
VA < 6/24 or partially sighted (1 statement of special needs)			**3**	**2**
VA < 6/18 (1 bilat. optic atrophy)			**2**	**1**

Based on age-category, 4/17 (24%), 4/58 (7%), and 8/110 (7%) with visual deficits 1-year post-diagnosis, were diagnosed between 1-6, 7-11, and 12-16 years respectively.