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Title Page

Sensory Processing Difficulties in Opsoclonus-myoclonus Syndrome: A pilot project of presentation and possible prevalence

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Abstract

Opsoclonus-myoclonus syndrome is a rare but serious neurological condition resulting in loss of control of eye movements, often accompanied by difficulties in posture and movement control with reports of sensory sensitivities potentially impacting on behaviour. This pilot study characterises the presence of atypical sensory behaviours in opsoclonus-myoclonus syndrome through questionnaire survey of a cohort of families. The Short Sensory Profile, Vineland Adaptive Behaviour Scale and Developmental Behaviour Checklist were distributed to 30 families; 16 were returned anonymously. Atypical sensory behaviours were identified in a large proportion (62.5%). Children reported as being more anxious showed greater sensitivity to auditory stimuli (U (14) 11, $p = .026$). This is consistent with recent recognition of more extensive disease neuro-cognitive effects in Opsoclonus-myoclonus syndrome. Further research is needed to increase understanding of the complex pathology of this disease and to provide indicators for sensory and behavioural as well as pharmacological interventions.

Key words: Opsoclonus-myoclonus syndrome, Sensory Processing Disorders, Anxiety
Words 147/150

Introduction

Opsoclonus-myoclonus syndrome, also known as 'dancing eye syndrome', typically presents in children between 12 months and 3 years of age with variable outcome.[1-4] Children develop opsoclonus, a rapid, involuntary, conjugate, multidirectional, saccadic eye movement disorder; accompanied by myoclonus and ataxia. Irritability and sleep disturbance are often also observed; and less frequently neurological symptoms like vomiting and mutism may also feature.[3,4] Whilst the illness may be monophasic or multiphasic; many (over 50%) children experience learning, behavioural and coordination difficulties with problems persisting into adulthood.[3,4,6,7] Recent report suggests some improvements in outcomes with increased immunosuppression[8].

A neuroblastoma is present in about 50% of children and presumed to be an immune-mediated para-neoplastic condition whereby immune perturbations have been identified in the form of reactivity of patient derived humoral and cellular components to tumour cell lines.[5] Central nervous system auto-reactivity in opsoclonus-myoclonus syndrome may also occur in the absence of tumour, where a post-infectious aetiology is purported to be the trigger.[3,4] Clinical and radiologic evidence now points to extra-cerebellar involvement in opsoclonus-myoclonus syndrome.[9] Electrophysiological studies also appear to support the clinical observation that some of the neuro-behavioural sequelae following opsoclonus-myoclonus syndrome may result from aberrant sensory processing and modulation.[10,11]

Sensory modulation dysfunction is defined as the impaired ability of an individual to regulate and organise responses to sensations in a graded and adaptive manner and appropriate to situational demands; manifesting as over-responsiveness (reduced sensory threshold); under-responsiveness (high sensory threshold); or sensory seeking behaviours which may be representative of an under-responsive sensory system.[12-13] It has been conceptually and empirically associated with a number of behavioural manifestations particularly amongst individuals with neurodevelopmental disorders[14-22] Atypical sensory processing has been suggested as a major factor reducing participation and engagement in daily activities across a number of developmental disorders.[12-23] Patient reports, clinical observations of features

associated with opsoclonus-myoclonus syndrome and electrophysiological evidence suggest that over-responsiveness to sensory stimuli may significantly impact on participation in daily activities.

The relative contribution of sensory over-responsiveness as an independent symptom is however confounded with the overlap of symptoms with those of anxiety and the lack of a 'gold standard' for the identification of sensory modulation dysfunction or sensory processing disorder, with questionnaire report most commonly used[12,22,23]. The determination of an abnormal behavioural response (a defined cut point) from quantitative analysis of deviations of what generally constitute a range of typical behaviours, may be augmented by biological measures (e.g. electrodermal responses) as developed by Miller and colleagues, however the correlation between these and behaviour report are not universal. [12,13]

This background provides a plausible argument for sensory modulation dysfunction and or anxiety affecting performance in daily activities for young people opsoclonus-myoclonus syndrome. The aim of this study was to gather preliminary information, via an anonymous questionnaire survey, of the presence of sensory processing problems (and/or anxiety) in opsoclonus-myoclonus syndrome and their impacts on cognitive, social and adaptive functioning, in order to guide future research.

Methods

This project was designed as a questionnaire survey of a convenience cohort of families with a young person with opsoclonus-myoclonus syndrome. Ethical approval conforming to the Declaration of Helsinki was received from the National Research Ethics Service (2/LO/0115).

Participants

Questionnaire packs with information regarding the project were distributed to families with a child/young adult with opsoclonus-myoclonus syndrome via the Dancing Eye Syndrome Support Trust, with an anonymous code linking the questionnaires within the pack. Stamped self-addressed envelopes were provided for return to the research team. Informed consent was assumed via return of completed questionnaires. Sample size was therefore limited to 30 registered families with a return rate of 53%

resulting in a sample of 16. Recruitment and enrolment did not distinguish between individuals at different disease phases (eg. Acute, on-going treatment or remission).

Measures

Demographic information was collected alongside questions regarding the age Opsoclonus-Myoclonus Syndrome was acquired and reason (if known) alongside any current medical treatments, therapeutic procedures received and educational placement.

The Short Sensory Profile Caregiver Questionnaire[24] was completed to establish the profile of sensory responses. Sensitivity and specificity have not been published although discriminate validity is reported as greater than 95%. Total scores range from 38 to 190 with cut-off scores defining typical (155-190) vs probable (140-154) and definitely atypical (38-141) sensory behaviours determined from the standardisation of the Short Sensory Profile.[24]

The Vineland Adaptive Behaviour Scale [25], was used as a standardised scale of adaptive behaviour and development obtained through parental/key care giver report across four domains of social, communication, daily living and motor skills. Questions relate to relevant skill areas and contribute to an overall adaptive behaviour score. Good reliability and validity are reported.[25].

The Developmental Behaviour Checklist [26] a 96 item parent report instrument of childhood behavioural disorders was used to measure anxiety. An anxiety score is derived by summing the 12 items of the anxiety subscale with high scores reflecting high anxiety (score range 0-24). Items linked to sensory behaviour are limited to two and data can be rerun removing these items.

Data Analysis

Statistical analyses, using the statistical package SPSS (v.19), were undertaken relevant to the original aims. Missing data were prorated if less than 10% of subscale was missing as an average for the section. Descriptive statistics explored the distribution, characteristics and results of the children across questionnaires. Primary outcomes considered the proportion of individuals who are reported to have

sensory processing problems. Secondary outcomes considered the relationship between anxiety and adaptive behaviour and participation in daily activities; controlling for additional factors which may influence engagement and participation through Spearman correlation analyses and between group comparisons of children with and without reported anxiety. Model assumptions were checked and parametric (ANOVA) and non-parametric (Chi^2 and Mann Whitney U) analysis methods used as reported.

Results

Sixteen of 30 (53%) questionnaires were returned for 13 females and three males. Mean age was 7 years 6 months (range 21-168 months) at time of questionnaire completion. Time since diagnosis ranged from 6 months to 12.4 years (*M* 6 years, *SD* = 4.5). Eleven children were reported to have had a neuroblastoma and two a known viral infection with three not providing information. At time of study nine children were not on medication, one on cyclosphosphamide and three on prednisolone (See Table 1 and Appendix 1 for participant characteristics). Two returned Vinelands required prorating on one or two scales and two were not possible to prorate.

<Table 1 approximately here>

There were no significant differences in developmental ability between older or younger children, contrasting impact of relatively acute versus chronic stage of opsoclonus-myoclonus, nor between males and females (See Table 1). However, all but one of the older children showed significant delays in adaptive behaviour with percentile scores of five or below as opposed to only two of the five younger children[1]. There were also no differences in age, time since diagnosis, and overall developmental ability (as measured on the Vineland) between children reported on the Developmental Behaviour Checklist as being anxious or not ($t(13) < 135$, $p > .05$ across all measures; See Table 2).

[1] Data were re-analysed without the outlier on the Vineland and differences between younger and older children were only evident on overall adaptive behaviour with older children reportedly more delayed for their age across domains (*p=.053).*

Younger children (50%) were more likely to be on medication than children over 6 years (20%), at questionnaire completion, as they are temporally closer to disease onset. Older children (>6 years; n=8) showed poorer overall development as reported on the Vinelands (U (14) 7.5, p =.029). Older children (90%) had also received more therapies such as physiotherapy, speech and language therapy and occupational therapy and additional educational support than younger children (30%).

There were eight children reported to have atypical sensory behaviours (50%), two probable (12.5%) and six (37.5%) considered to be within the typical range according to the criteria of the Sensory Profile. Differences in sensory processing were evident between children reported as having anxiety and those that did not with children with anxiety reported to have hyper-responsiveness to auditory stimuli (Auditory: U 11, p =.026, r = .593) which contributed to the differences in the Short Sensory Profile total ($\underline{U}$ 12.5, p = .038, r = .513). The numbers of children reported as having anxiety were significantly higher amongst children who also met criteria for atypical sensory behaviour (χ^2 (2) 8.56, p = 0.032) with the likelihood ratio reducing (χ^2 (2) 5.74, p = 0.06) with removal of the two children who were below the standardisation age (3 years) of the Sensory Profile and Developmental Behaviour Checklist. See Table 2 and Figures 1 and 2.

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Significant correlations were evident between sensory behaviours and anxiety subscale of Developmental Behaviour Checklist (*rho*= -.724, p = .028), standard scores and percentile scores of the Vineland *(rho*=.761, p = .002, *rho* = .593, p = .025 respectively) and the anxiety scale of the Developmental Behaviour Checklist and both Vineland standard scores and percentiles (*rho* = -0.708, p = .033; *rho* = -0.681, p=0.043). In view of two children being outside of the standardisation range on the Sensory Profile and Developmental Behaviour Checklist we reanalysed our data without these children with similar trends across results.

Discussion

This study aimed to gather preliminary information via questionnaire survey of the potential presence of sensory processing problems in opsoclonus-myoclonus syndrome and possible impact on cognitive, social and adaptive functioning. . Our results suggest that anxiety was associated more specifically with hyper-sensitivity to auditory stimuli rather than a generalised sensory modulation dysfunction in some, but not in all of the children.

Ben-Sasson and colleagues[16], in a meta-analysis of sensory modulation dysfunction, in individuals with autism spectrum disorders, showed that presentation of sensory modulation dysfunction is influenced by age, severity of autism and type of control group used for comparison, while Lane et al[14] found anxiety to be influenced by the magnitude of response to sensory stimuli. Sensory-based phenotypes based on specificity of sensory responsiveness in autism have also been suggested.[27] These findings suggest a potentially important interaction between anxietyand expression of sensory symptoms. Schneider and colleagues[28] have used a primate model to show the influence of prenatal stress (including exposure to alcohol and or adverse sounds) on both striatal dopamine levels, evaluated using positron emission tomography, and increased withdrawal to sensory stimuli (aversion to repetitive tactile stimuli and reduced habituation to stimuli across trials in 5 to 7 year old rhesus monkeys). Infant stress (neuroinflammation) may therefore be hypothesised to contribute to an attenuation of sensory sensitivity, as a protective response in the initial stages but which may later contribute to presentation of negative behaviours (e.g. withdrawal from light touch or auditory stimuli). What is unusual in our data is that this negative response appears to be relatively restricted to auditory stimuli.

We had anticipated that more atypical responses would be evident across sensory systems in line with Schneider et al.[27] as a systemic response to infant stress. Currently, the selectivity of the hypersensitivity to the auditory system in patients in this cohort is surprising as opsoclonus-myoclonus syndrome appears to involve oculomotor components including blurred and or double-vision. A recent neuroimaging study has however suggested more extensive degeneration with cortical thinning across visual as well as

cognitive, language, and motor areas additional to cerebellar involvement of pontine-cerebellar regions.[9] What has not previously been considered is whether there is a direct effect on the inferior colliculus and the processing of auditory stimuli. Autoimmune mechanisms may therefore be more 'site specific' in opsoclonus-myoclonus syndrome than originally conceptualised.[3,4,29]. What is not possible to conjecture from our results is the representation of atypical sensory behaviours in opsoclonus-myoclonus syndrome based on aetiology as evidenced in developmental and genetic disorders.[27,30-32] . As such, the potential selective impact on auditory processing in opsoclonus-myoclonus syndrome could perhaps be explained by the focality of inflammation around inferior colliculi and auditory pathways or the vulnerability of auditory processing to a more generic disease effect (cortical or sub-cortical) with consequent developmental impact.

There is as yet no known relationship between hyperacusis and auditory sensitivity across different environmental sounds. The complex pathway of opsoclonus-myoclonus syndrome with some children showing rapid spontaneous improvement without treatment and others showing persistent presentation of multiple symptoms with unclear links to aetiology suggests a complex interaction between the infant, disease and environment.[4] Of note is the potential influence of prednisolone on behaviour. Analysis of the reported behaviours of the three children in our study on prednisolone (ages 1year 9months, 4years 2months and 8years 9months), showed all to be reported as anxious, but all within normative ranges on the Vinelands and only one showing significant sensory behaviours on the Sensory Profile. Consideration of the interaction of medication on symptom expression in opsoclonus-myoclonus syndrome requires elucidation through further research.

While modest, our results suggest a potential role of auditory hypersensitivity in opsoclonus-myoclonus syndrome which may be integral to the condition and or a secondary consequence in some cases, which may exacerbate or represent the persistence of symptoms. What remains unknown is whether sensory modulation dysfunction is evident in other neuroinflammatory diseases which might provide potential markers for the disease process. Comparisons with other diagnostic groups that have received immunomodulation as well as those with neurodevelopmental disorders with and without

sensory processing deficits, may provide insights into this complex disease, neurological sequelae and current management. The important aspect of the current findings is the identification of a risk of sensory processing difficulties, particularly of auditory stimuli, linked to anxiety in opsoclonus-myoclonus syndrome. We recommend that parents and clinicians consider the child's responses to auditory stimuli through observation, and report of behaviours across contexts with consideration of appropriate therapies to reduce sensory sensitivity.

Limitations

The use of the Short Sensory Profile to identify sensory processing disorders in children is confounded by the limited discrepancy between chronological age and mental age and lack of specificity due to overlay with diagnostic criteria within the Sensory Profile . The anonymous distribution of questionnaires in this survey did not allow for provision of age specific questionnaires nor exploration of aetiological and early medical inventions as factors contributing to outcome. While variations of the full Sensory Profile exist for infants and adolescents, the Short Sensory Profile is only available in one version which may not be sensitive to differences in younger and older children – although this limitation is more likely to magnify results. The disproportionate response rate with more females than males is not considered to represent a bias of presentation of opsoclonus-myoclonus syndrome in either the acute or chronic phases. However, gender differences in the persistence of behaviour symptoms remains an unanswered question and further research with a broader population is needed. While our sample size was small, the recruitment rate was none the less high for this type of study (53%). This raises the question as to whether families may have been invested in the concept of sensory processing difficulties in opsoclonus-myoclonus syndrome. The study could also have been improved with the use of a sibling control group, however this would have increased the burden to respondent families.

Conclusion

The atypical responses to sensory processing in a large proportion of our modest sample support the need for further research in this area to: a) increase understanding of the complex pathology and developmental, psychological and sensory functioning of opsoclonus-myoclonus syndrome and their interactions and b) provide indicators for behavioural as well as pharmacological interventions. Findings will provide invaluable markers regarding the need and nature of more structured and sophisticated projects.

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Author Contributions

DG was the principal investigator; involved in all aspects of the study including design, ethical preparation and submission, data collection and analysis and overall responsibility for manuscript preparation

JT was involved in design of the study proposal and assisted with the paperwork for ethical preparation and submission, execution of the study and manuscript preparation.

KP was involved in study execution, research governance, and review of data and manuscript preparation.

ML and BL were involved in data analysis and review and manuscript preparation.

Declaration of Conflicting Interests

The authors declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article. All anonymised data is available for scrutiny from the first author.

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Ethical Approval

The study was approved through the UK Integrated Research Application System of London and Surrey Borders (Ref: 12/LO/0115) and conducted in accordance with the Declaration of Helsinki.

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Figure legend

Figure 1 Short Sensory Profile domain scores of children with and without anxiety as reported on the Developmental Behavior Checklist

Figure 2 Numbers of children meeting criteria for sensory processing disorder with or without anxiety

Table 1. Participant characteristics by younger and older age groups

Group	Age in months at time of study Mean (SD) range	Time (months) since diagnosis Mean (SD) range	Gender	VABS Percentile Mean (SD) range
6 years or less N=8	44.1 (23.2) 21-81	26.1 (22.2) 6-67	5 females, 3 males	13.7 (7.3) 4-21
7 years or older N=8	141.6(20.8)105-168	119.6 (29.4)63-147	8 females	13.3 (31.0)1-5(90[a])
Total n=16	92.8 (54.7) 21-168	72.9 (54.5) 6-147	13 females,3 males	13.4 (23.2) 1-90

[a] One outlier with a percentile score of 90; Abbreviation: VABS, Vineland Adaptive Behaviour Scale

Table 2. Presentation of Behavior and Sensory Profile by presence of Anxiety

Group (n)	Age months Mean (SD) range	Time (months) since diagnosis Mean (SD) range	VABS Percentile Mean (SD) range[a]	SSP Total score Mean (median) range
No Anxiety (8)	81.5 (54.8)	61.0 (55.0)	21.9 (31.1)	162.6 (164.5)
	21-146	6-133	2-90	133-184
Anxiety (8)	104.3 (55.7)	84.8 (54.9)	5.0 (6.0)	132.7 (137.5)
	31-168	11-147	1-18	89-174
Total (16)	92.88 (54.7)	72.9 (54.5)	13.4 (23.2)	147.6 (149.5)
	21-168	6-147	1-90	89-184

[a] n = 7 per group; Abbreviations: VABS,Vineland Adaptive Behavior Scale; SSP,Short Sensory Profile