J Inherit Metab Dis (2009) 32:353–360
DOI 10.1007/s10545-009-1149-1

SYMPOSIUM ON NEUROTRANSMITTER DISORDERS

Sensory integration intervention: Historical concepts, treatment strategies and clinical experiences in three patients with succinic semialdehyde dehydrogenase (SSADH) deficiency

S. V. Kratz

Received: 25 January 2009 / Submitted in revised form: 25 January 2009 / Accepted: 2 March 2009 / Published online: 20 April 2009

Summary This paper is a review of clinical experiences providing developmental therapy services for three boys diagnosed with paediatric neurotransmitter disease. The clinical presentation of paediatric neurotransmitter diseases might parallel other diagnostic characteristics seen in a typical paediatric therapy clinic (i.e. hypotonia, motor and cognitive delays, coordination, expressive speech, and ocular motor difficulties.) From the clinical perspective of the author, sensory integrative function is but one aspect of a thorough evaluation and treatment plan for all patients. The manifestations of sensory integration dysfunction (SID), also known as sensory processing dysfunction (SPD), can occur alone or be concurrent with a variety of known medical, behavioural and neurological diagnoses. These manifestations of SPD can include, but are not limited to: hypotonia, hyperactivity, irritability, distractibility, attention difficulties, learning difficulties, clumsiness and incoordination, instability, poor motor skills, social-emotional difficulties, and behavioural problems. This paper summarizes the theory and practice applications of sensory integration. The author discusses clinical experiences providing occupational therapy services utilizing sensory integration methods and strategies with clients who were eventually diagnosed with SSADH deficiency.

Abbreviations

PND	paediatric neurotransmitter disease
SI	sensory integration
SIPT	Sensory Integration and Praxis Tests
SPD	sensory processing dysfunction
SSADH	succinic semialdehyde dehydrogenase

Introduction

This paper is a review of clinical experiences providing developmental therapy services for three boys diagnosed with paediatric neurotransmitter disease (PND). The clinical presentation of PNDs might parallel other diagnostic characteristics seen in a typical paediatric therapy clinic (i.e., hypotonia with resulting balance and posture deficits, motor and cognitive delays, incoordination, expressive speech deficits, and ocular motor difficulties.) From the clinical perspective of the author, sensory integrative function is but one aspect of a thorough occupational therapy evaluation and treatment plan. The manifestations of sensory integration dysfunction (SID), also known as sensory processing dysfunction (SPD), can occur alone or be concurrent within a variety of known medical, behavioural and neurological diagnoses. These manifestations of SPD can include, but are not limited to: hypotonia, hyperactivity, irritability, distractibility, attention difficulties, learning difficulties, clumsiness

Communicating editor: Michael Gibson

Competing interests: None declared

Presented at the 2nd Pediatric Neurotransmitter Disease (PND) Association Symposium, 'Medical Management of Pediatric Neurotransmitter Disorders: A Multidisciplinary Approach', 18–19 July 2008, Hyatt Dulles Hotel, Herndon, VA, USA.

S. V. Kratz (✉)
Special Therapies, Inc.,
W238N1690 Rockwood Drive, #500,
Waukesha, WI 53188, USA
e-mail: specialtherapies238@yahoo.com

and incoordination, instability, ocular motor dysfunction, poor motor skills, social-emotional difficulties, and behavioural problems. This paper summarizes the theory and practical applications of sensory integration. The author discusses her experience providing occupational therapy services utilizing sensory integration methods and strategies with clients who were eventually diagnosed with SSADH deficiency.

History of sensory integration

Sensory integration is an innate neurobiological process that refers to the interpretation of information about the environment received through the senses. A. Jean Ayres, an occupational therapist and educational psychologist, offered a definition of sensory integration as she observed human occupation beyond the idea of intricate synaptic connections typical of neuroscience. Her definition, the 'organization of sensation for use' brought forth a fusion of neurobehavioural insights with practical solutions to dysfunction in young children who are affected by an inefficient nervous system (Ayres 1972, 1989).

Sensory integration then evolved into a clinical specialty area in occupational therapy that may be integrated into occupation-centred practice (Fisher and Murray 2002). Ayres' work dating back four decades has evolved into the most rigorously researched aspect of occupational therapy practice. The term further gives a frame of reference that evolves with neuroscience findings, for understanding how the brain organizes sensations for engagement in occupation (purposeful activities that give the individual meaning).

Ayres developed the theoretical constructs, established standardized testing methods, and established an extensive clinical approach (Ayres 1972). The Sensory Integration and Praxis Tests (SIPT) assist in the identification and classification of subtypes of sensory integration deficits in children (Ayres 1989).

Activity analysis is the art and science combination of selecting a therapeutic environment and use of medium to meet therapy objectives that address these identified sensory processing (and functional) deficits. The occupational therapist utilizing a sensory integrative approach designs the environment in a way that the individual can gain skill mastery, tapped by their own inner drive, towards more purposeful engagement and raise performance levels within meaningful tasks (Bundy 2002).

The ability to produce adaptive responses reflects sensory integration and reflects the child's purposeful use of sensory channels. Some examples of adaptive responses include motor control, coordination, quality of movements, social and physical interactions, organization of self and behaviour, flexibility of thought and actions, ability to modulate self from sensory experiences and execute a well-matched reciprocal action, and language use (Ayres 1972, 1989). Sensory integration gives occupational therapists a practical, valid, and reliable model in which to interpret and measure a person's perception and meaning of sensory inputs of the vestibular, proprioception and touch systems.

Sensory processing dysfunction

The theoretical frame of reference of sensory processing function thus holds a theoretical link to motor coordination, language comprehension and usage, perceptual motor skills, attention and learning, emotional stability and behavioural control and self-control (Ayres 1972, 1989). Sensory processing dysfunction has been associated with sensory deprivation of early childhood, fragile X, autism spectrum, ADHD, brain injuries and fetal alcohol syndrome (Franklin et al 2008; Miller et al 2001). Clinically, sensory processing dysfunction is also associated with cerebral insults, seizure disorders, and other neurological conditions. Primate models have been firmly established for sensory processing dysfunction related to prenatal stress. A growing body of research exists on a nonhuman primate model to examine the neurochemical and developmental basis for sensory processing disorders (Schneider et al 2007, 2008). It is a reasonable assumption that children identified in the emerging discoveries of paediatric neurotransmitter diseases might also have difficulty adaptively responding to the sensory input of their environment.

Assessment

Careful assessment is necessary to identify whether sensory processing dysfunction is a factor influencing the presenting problems. Whenever possible, the trained therapist will utilize standardized testing and include scores from other sources of information, such as extensive parent interviews about daily life and struggles, interviews with other caregivers or teachers, systematic clinical observations, and behavioural checklists that are researched under the sensory integration model. The therapist synthesizes the data from all available sources to form a hypothesis of what sensory systems are likely a

contributing factor in the presenting functional deficits (Parham and Mailloux 2001).

Clinical evidence and empirical research

Most existing research on the use of sensory integration methods focuses on young children with a variety of developmental and learning difficulties, including but not limited to developmental coordination disorder, learning disabilities, attention disorders, behaviour problems, autism, and other categories (Baranek 2002, Miller et al 2001, Smith Roley et al 2001). Documenting the effectiveness of occupational therapy using a sensory processing approach is a challenging endeavour, though over the course of several decades many research studies have investigated clinical effectiveness of sensory integration therapy (Daems 1994; Mulligan 2003a, b). There have been many studies assessing the effectiveness of 'sensory-based' interventions or specific isolated strategies. Sensory-based interventions that reference Ayres' original works lack in differentiation and clarity of terms. The results from earlier studies were encouraging, but as methodologies have become more rigorous the results have been less than favourable. One study using meta-analysis confirms this finding (Vargas and Camilli 1999).

Parents of children with autism, a broad spectrum of neurobehavioural dysfunctions, are increasingly turning to sensory integration treatment to help their children deal with the disorder. In a survey study of 112 families, 71% of parents of children with autism who pursued complementary and alternative to traditional treatments participated in sensory integration methods, and 91% found these methods helpful (Hanson et al 2007). A new clinical study found that children diagnosed with autistic spectrum disorders who underwent sensory integration therapy exhibited fewer autistic mannerisms compared with children who received standard treatments (Pfeiffer and Kinnealey 2008). The author's experience is that many of the 'soft signs' of autism, for instance, dyspraxia, difficulty with motor initiation and control, and poor functional use of proprioception, reflect inadequate sensory processing. Autism has been noted to be a differential diagnosis in the assessment of paediatric neurotransmitter disease, as is the case of one of the boys in this review.

Identifiable symptoms of neurotransmitter disease include hypotonia, gross and fine motor deficits, and cognitive delays, yet these are also common functional problems in other disorders such as Down syndrome and cerebral palsy. These symptoms frequently necessitate referrals for physical medicine intervention. Uyanik compared the effects of sensory integrative therapy alone, vestibular stimulation in addition to sensory integrative therapy, and neurodevelopmental therapy, on 45 children with Down syndrome. The results suggested that sensory integration treatment, vestibular stimulation and neurodevelopmental therapy were effective in children with Down syndrome (Uyanik et al 2003). In another study, sensory integration treatment was provided to random controlled groups of children with spastic diplegia. Results indicated that both group and individual treatments had a measurable effect that was consistently greater than that of controls (Bumin and Kayihan 2001).

Mulligan examined the evidence for occupational therapy using a sensory integration framework with children (Mulligan 2003a, b). Schaaf and Miller review the state of the current evidence to support this treatment approach, suggesting that consensual knowledge and empirical research are needed to further support its utilization for a variety of developmental disabilities. This is especially critical given the public pressure by parents of children with disabilities to obtain the most effective services, and who have anecdotally noted the utility of sensory integration therapy for helping their children function more independently (Schaaf and Miller 2005).

A review of the effectiveness research identified few studies that reflected true adherence to the core principles of sensory integration intervention (Parham et al 2007). There exists very little research that supports the effectiveness of any intervention for children with chronic or mild disabilities across all disciplines (Parham et al 2007; Ottenbacher 2002). In spite of considerable effort to investigate sensory integration treatment, few studies have truly accomplished the task of achieving rigorous study design. Furthermore, there is no evidence that indicates that sensory integration intervention is more or less effective than other interventions. With the emerging field of study in paediatric neurotransmitter diseases, this challenge only intensifies.

Core concepts of treatment

Sensory integration treatment differs from other neurodevelopmental theoretical models or modalities. A key difference capitalizes on the child's intrinsic motivation as a catalyst in the sessions. In order to set up the environment, the therapist uses a wide range of play media, activities, and/or equipment that stimulate specific sensory receptors to promote integration of

the sensations for a result in the production of adaptive responses. Adaptive responses can be reflex integration, adequate postural adjustments, movement successes, increased alertness and awareness of input, self-regulation without maladaptive emotional reactions, and abilities to start and participate in the task through to completion. The adaptive responses a child demonstrates are unique and individualized. What might be adaptive to one child might not be for another. The outcome measure is not in simply having the child execute a target response; rather the objective is to help the child reach a change in the complexity of the adaptive response that moves them towards a higher level of independence (Bundy 2002).

Every encounter with a sensory agent can have either a facilitatory or an inhibitory effect on the child's nervous system, and it is the therapist's role to set up the target parameters. Furthermore, the therapist recognizes whether facilitation or inhibition occurred through analysis of the adaptive responses of the child. The clinical reasoning of the therapist creates the design of even the simplest activity for independent success. This term is called 'scaffolding' the session. To the untrained eye, therapy appears to be merely playing with the child, but a well-executed treatment session means that the therapist is working very hard throughout to have that 'play' improve the child's nervous system processes (Ayres 2004).

The vestibular system and the information it processes are often a significant aspect of treatment planning. Movement in rotation is carefully utilized to stimulate the semi-circular canals and is believed to facilitate phasic muscle tone. The goals of improving alertness and awareness of three-dimensional sense of space are related to various orbital, axial, and arcing planes of movement. The three planes of direction that exist with the canals means that horizontal, vertical, and angular planes of movement are factored into the activities at appropriate times. Linear acceleration (moving forward in a straight plane), the use of gravity (often by incorporating head inversion), and vibrations are utilized to stimulate the otoliths. Otoliths are involved in facilitation of tonic muscles that contribute to balance and posture by increasing awareness to gravity and activating anti-gravity muscles. Stimulation of the utricles is believed to assist in ocular motor activation; and stimulation of the saccules is believed to specifically facilitate a postural response.

Swings are the most common equipment used to incorporate vestibular input. Additionally, objects on the ground such as a large air mattress with an unstable base, or rubber tyres that can be sat, stood, or bounced on, or suspended, can provide the desired input. Sitting on a suspended inner tube not only offers the vestibular options, but this task can stimulate Golgi tendons and muscle spindles to fire proprioceptive input.

Vestibular input is an exceptionally powerful input, especially when the proprioception and tactile systems are not helping to organize the motor systems. Delayed reactions might occur in this situation, which can actually disorganize that system. The therapist must be a keen observer of every physiological cue and intervene at the earliest sign of physiological stress. The therapist must recognize when the task has gone beyond the realm of the 'just right' challenge and make modifications that return the task to that realm. Vestibular processing is believed to help unify the other systems (Ayres 1972, 1989).

To stimulate somatosensory pathways of proprioception and deep touch, Golgi tendon organs and muscle spindles are activated through joint compression or traction, muscular contractions, and full-body compression. Active proprioceptive input in functional tasks is accomplished through heavy work patterns of pushing, pulling, and carrying. Passive input (i.e., wrapped in a compression garment) is believed to stimulate deep touch pathways. Proprioception and deep touch pressure are hypothesized to be organizing to support and improve: attention and arousal levels, body awareness and position in space, and muscle tone, and to modulate sensory perceptions. Equipment that is commonly used includes trapeze bars, large foam pillows or mats, elastic fabric hammocks, together with moving against resistance through tight-fitting spaces.

Touch activities are designed to stimulate the complex layers of tactile reception. Goals include increase localization of touch, visually directed hand movements through tactile, discrete control of tools, discrimination of textures, and emotional tolerance for various inputs. Light touch tends to facilitate arousal levels, awareness and attention. Conversely, deep pressure touch input tends to inhibit and modulate attention and emotions, but can also facilitate postural stability, joint awareness, and muscle contractions.

Endless arrays of media can be used, including fabric swings, use of sand or grain for digging in, play dough, and finger painting with paints, pudding, and shaving cream. If touch is alarming, it is commonly effective to stimulate deeper touch input and proprioception to normalize the responses to light touch. If writing is a problem, it is a sensory integration strategy to first treat improving tolerance and discrimination of various tactile media, rather than 'practising' the frustrating tasks using paper and pencil. Examples might be to shape letters with bread dough, or use sticks in wet sand, or rub chalk over sand paper.

Ocular motor coordination is a direct reflection of sensory integration. Initially reflexive in nature, ocular responses to vestibular input and postural changes prepare the eyes for movement in skilled pursuits and focusing. Primary ocular motor skills of tracking, convergence, divergence and gaze shifts embedded into functional tasks are a reflection of the dyadic integration between vestibular input and the proprioceptive reactions from the ocular motor muscles. Bilateral coordination of ocular muscle groups may parallel the degree to which the extremities are coordinated in timing and rhythm.

Not to be overlooked, the oral structures can have the same challenges of sensory awareness, apraxia, and motor control. An important factor in speech production is the sensory processing of extra- and intra-oral structures. Light touch awareness and deep pressure processing are significant factors in the adaptive use of tongue, lips, jaw, swallowing mechanisms, and neck stability. Various blowing toys, games involving exhalation, and masseter pressure to develop discriminative bite force are a few examples of clinical application.

Skills of praxis can be described as purposeful and meaningful use of proprioception and touch in automatic reactions, especially measured in performance of novel events. Praxis components include ideation, planning, execution, following sequential steps, organization of motor responses, and constructional skills. Praxis is the ability to translate an idea into action; the act of implementing that idea; and completing all sequential steps of the task. Praxis is how competencies are built and how higher levels of independence are mastered. In the treatment process, the therapist is challenging the child to choose, plan, set up, and follow sequential steps of the activities as a way of promoting praxis skills.

Treatment activities can address specific problems directly, such as postural and ocular control. More often, however, treatment activities address general function indirectly. A child may not be able to put on socks independently because they cannot feel the tension necessary to stretch the garment, nor adjust the fabric, owing to poor tactile awareness. Treatment activities probably would not include repeated training of the act of putting socks on, especially if frustration and a sense of failure exist. Instead, tasks such as pushing the feet into a tub of popcorn to feel for hidden sponge toys, or shaping clay around hands and feet for a variety of texture and resistance experiences, or even imaginary painting the skin with various brushes in play might be used to increased tactile awareness and discrete discrimination. In an actual experience, one of the boys in this review was able to independently start and finish the donning of socks following three such treatment sessions.

Presenting symptoms of clients with SSADH deficiency

In a private suburban therapy clinic in the Midwest, three boys diagnosed with SSADH deficiency and their families sought specialized services that specifically included occupational therapy for sensory integration, and speech therapy that also embraced this theoretical approach. All three patients had been involved in intensive Birth-Three programming and beyond. All had had the opportunity to experience a variety of philosophical approaches to physical medicine therapy in different settings. All three boys' therapy programmes were well established prior to the onset of services with a sensory integration focus. Initial symptoms are reviewed retrospectively for each child.

Child 1: Male ('John'). Onset of symptoms was recognized in infancy. He received an accurate diagnosis of SSADH deficiency at age 18 months. Functional deficits included:

- Severe hypotonia despite independent ambulation.
- Poor co-contractions in weight bearing, poor postural and balance reactions.
- Upper extremity function impaired by lack of shoulder (proximal) stability with significant fine motor deficits.
- Ocular motor control for visual guidance of hands in tasks.
- Lack of body control required years of supervision and adult assistance for all daily living tasks to ensure safety.
- Poor postural and functional use of proprioception increased his risk of injury with a lack of perceptual-motor judgement and problem-solving abilities.
- Significant ocular motor control and poor visual tracking abilities.
- Severe apraxia of speech and dysarthria
- All daily living skills required maximal assistance.

Child 2: Male ('Jordan'). Onset of symptoms was in early infancy. SSADH deficiency diagnosis was received at 12 months of age. Functional deficits included:

- Severe hypotonia as well as a lack of voluntary muscular contractions for activity.
- He was non-ambulatory and required use of wheelchair, though he could crawl and roll.

- He required specific feeding therapy intervention by a swallowing specialist in addition to other therapy regimes.
- Severe deficits in fine motor development.
- He was essentially non-verbal owing to severe oral apraxia of speech.
- All daily living activities required maximal assistance to complete.

Child 3: Male ('Max'). Onset of symptoms was not apparent prior to diagnosis. His diagnosis of SSADH deficiency was obtained at age 1 month through genetic testing in conjunction with his older brother's workup. Functional deficits noted at age 4 years, when our clinic became involved, included:

- Moderate hypotonia was present.
- Ankle orthoses were required for gait assistance, though ambulation was independent but tenuous.
- Gravitational insecurity with concurrent intolerance to imposed movement
- Delayed postural and balance mechanisms interfering with motor skill mastery.
- Moderate to severe oral apraxia of speech
- He required moderate assistance with activities of daily living

Response to treatment

All three boys received private therapies (occupational and speech therapy) as well as local school-based level of intervention for physical, occupational, and speech therapy. The two brothers also received traditional physical medicine in a hospital setting for a while. The private therapy plan utilized sensory integration strategies along with neurodevelopmental therapy strategies, environmental modifications, adaptive equipment, and family education into theoretical principles and home programme activity plans. The sensory integration theoretical model was primary, but in combination, the sensory integrative strategies assisted their work towards more and more competencies in motivating activities, as well as subjectively observed improvements in muscle tone, postural control, midline stability, and extremity stability. Over time, these foundational improvements appeared to be underpinnings of generalizations to other areas of functioning: self-care skills, fine motor skills, attention and concentration, speech control. All three have gained measurably in movement abilities, increased sensory awareness, increased tolerance for movements, and increased cognitive competencies. All three boys have gained significant levels in their developmental profile and occupational competence and their progress remains dynamic and steady. All the boys are fortunate in having family members consistently dedicated to their care and progress, and have had financial resources to allow for reimbursement for these services.

This paper is not able to identify any single factor that might be identified as the agent of change, but rather to retrospectively review the sensory integrative approach that these two families sought and that they feel contributed to a greater rate and amount of progress.

Synopsis of a treatment process

The following description reflects the depth in which both activity analysis and therapeutic structuring are involved in the sensory integration treatment process. 'Max' presented with gravitational insecurity and intolerance for movement was a primary indication for intervention. He lacked adequate postural control and protective reactions under a hypotonic muscle base. He wore ankle-foot orthoses due to the hypotonia. These factors appeared to contribute to a child who did not spend much time exploring his environment or engaging in active learning opportunities of his own body potentials, and was basically avoiding movement activities.

A process of gentle guidance into a series of engagements with moving equipment was suggested for him in a series of scaffolding sequence. For several months this child was only comfortable watching other children ride a large platform swing, move along the floor on a scooter, or climb a 4-foot (1.3 m) loft and jump into a pile of pillows. Often, if encouraged to join in, he would hide and seek comfort from his mother or even leave the room. Activities were never forced so as to avoid alarming stress reactions and the risk of eroding his trust in the environment. One day he felt comfortable making a tentative attempt at pushing another child on a swing. Then another day he was comfortable climbing on the lower steps of the loft. Then he was able to build confidence to climb up the loft with his mother, but he fearfully clung to the wall and avoided the edge. Several times at this level of comfort he would climb up only to need complete help getting down the stairs doing a backward crawl.

Eventually one day, with his mother's help, he was able to be passively lowered off the loft into the pillows. The next week he was able to independently attempt a 'jump' into the pillows under his own control. Soon he was demonstrating more comfort when engaging with the equipment and began to initiate more with swing activities. His successes and motor competencies improved dramatically from that point of overcoming

his fear of movement. The eventual successful jump and landing into pillows was evidence of his gains in postural control and praxis as well as improved expression of vestibular and proprioception processing.

The scaffolding of this scenario included slow linear vestibular input on the swings while challenging his postural control until he could tolerate a variety of speeds and directions. The climbing and jumping activity was designed with a multitude of proprioception opportunities (which is hypothesized to modulate the vestibular and the limbic reaction of fear to movement). The floor in the treatment room requires walking on a variety of foam mats with differing densities and recoil. The steps to the loft are carpeted with thick threads for increased input into soles of feet to challenge balance reactions. The loft itself is a sturdy wood structure but also carpeted for additional tactile input as the child climbs, navigates and prepares a motor action.

There are three or more ways to ascend or descend this loft, each with its own design for deep pressure input—a slide, a rock climbing ramp, and various rope ladders or swings off which to exit. Sometimes new ways to ascend, descend and navigate this loft are created, for instance, using a long sturdy tunnel to make a bridge as an 'escape hatch'. The therapy is not in just navigating the crawling or sliding through the tunnel, but in encouraging the child to secure it by tying ropes or by anchoring with pillows and other supports. In other words, 'build your own escape hatch'. The proprioceptive 'lesson' thus is to feel the weight of the tunnel, feel how much force is needed to secure it, and feel when the tunnel is actually secure before attempting a crossing. This is an example of how higher cortical functions (problem solving, error recognition, and judgement) are added into the therapeutic process and help to advance the complexity of the adaptive responses.

There was marked and sustained improvements in areas of mobility, endurance, coordination, engagement, and social-emotional competencies in this child following the day he successfully motor-planned the 'jump' and masterfully landed in the pillows. His verbalizations increased exponentially. His frequent lethargy in the clinic reduced dramatically and he now begs his mother and therapist to 'play in the big room'. His social timidity seemed to disappear and he emerged as quite a vocal social being. Not only did he accept more motor and balance challenges on offered equipment, he started to seek out the environment on his own through movement exploration. He changed from a person who avoided movement and was impaired by hypotonia into a boy eagerly anticipating and investing into his physical environment. His level of hypotonia subjectively changed towards the normal range.

Parents' perspective on sensory integration treatment

Each mother provided her perspective when asked to summarize their interpretation of sensory integration treatment. The first mother stated: 'I could describe what sensory integration did for my son. It helped glue him together. It helped with his fine and gross motor skills as well as his balance. It was hard for 'John' to get his appendages to do what he wanted them to do. Before we tried sensory integration, we tried a handwriting programme, along with many other therapies. It was excruciating for him. After the sensory integration work, he is now capable of writing lists, short messages, numbers and his name. He also types short stories on a keyboard. Likewise, some days he couldn't walk through a door without hitting one side of it. I think sensory integration contributed to 'John's' improved balance and eventually him being able to ride a two-wheel bike.'

The mother of the two other boys stated: 'Sensory integration gave us a clearer direction and understanding of both my sons' difficulties with movement. That helped a good deal just to have a perspective of why things were so hard for them to do themselves. The most important things that helped were the activities and practical solutions to work on the problems; and at the same time these activities were highly motivating and fun to do.'

Discussion

It is a reasonable assumption that in the presence of neurotransmitter disease, the reception and processing of sensations through some or all channels would likely be adversely affected. Furthermore, the synthesis of information in higher cortical areas, the adaptive responses and ultimate 'meaning' of these sensations may also be impaired. As research advances a collective understanding of neurotransmitters' complex function, occupational therapists should continue to explore how this knowledge translates into human occupation performance and choose effective treatment approaches that best serve this emerging and complex population.

Physicians who distinguish and treat patients with paediatric neurotransmitter disease recognize the value of recommending physical, occupational and/or speech therapies. Therapists have long been exploring effective methods to remediate the functional problems

presented in clinic, regardless of diagnosis. It is fair to assume that many children who have reached a diagnosis of PND are involved in a local therapy programme(s) if services are available and accessible.

This retrospective review briefly summarizes how theoretical principles of sensory integration were embedded into the scope of practice, clinical reasoning, and treatment planning of three boys with the diagnosis of SSADH deficiency. Implications for clinical practice are that the lack of empirical data prevents a specific endorsement of sensory integration therapy. However, the existing validity of the sensory integration approach offers therapists a valuable option for training and preparing to work in the intervention of a variety of neurological disorders (American Occupational Therapy Association 2008).

Acknowledgements The author wishes to acknowledge the Pediatric Neurotransmitter Disease Association, National Institute of Neurological Disorders and Stroke, and Office of Rare Diseases (ORD) at the National Institutes of Health (NIH) and The Johns Hopkins School of Medicine. This paper complies with the requests of NIH Grant # 1R13NS060363–01.

The author also wishes to acknowledge with deepest gratitude, the countless clients and their families who continue to graciously teach us how to fuse theory into practice within the clinic and community settings.

The Symposium was supported in part by R13 NS 60363 from the NIH NINDS and Office of Rare Diseases (ORD), and the Johns Hopkins University School of Medicine.

References

American Occupational Therapy Association (2008) *The Guide to Occupational Therapy Practice*, 2nd edn. Occupational Therapy Practice Framework Practice Guidelines – Complete Set. Bethesda, MD: American Occupational Therapy Association.

Ayres AJ (1972) *Sensory Integration and Learning Disorders*. Los Angeles: Western Psychological Services.

Ayres AJ (1989) *Sensory Integration and Praxis Tests*. Los Angeles: Western Psychological Services.

Ayres AJ (2004) *Sensory Integration and the Child: Understanding Hidden Sensory Challenges*. Los Angeles: Western Psychological Publishers.

Baranek GT (2002) Efficacy of sensory and motor interventions for children with autism. *J Autism Dev Disord* **32**: 397–422. doi:10.1023/A:1020541906063.

Bumin G, Kayihan H (2001) Effectiveness of two different sensory-integration programmes for children with spastic diplegic cerebral palsy. *Disabil Rehabil* **23**(9): 394–399. doi:10.1080/09638280010008843.

Bundy AC (2002) Play theory and sensory integration. In: Bundy AC, Lane SJ, Fisher AG, Murray EA, eds. *Sensory Integration: Theory and Practice*. Philadelphia: FA Davis Company.

Daems J, ed. (1994) *Reviews of Research in Sensory Integration.* Torrance, CA: Sensory Integration International.

Fisher AG, Murray EA (2002) Introduction to sensory integration theory. In: Bundy AC, Lane SJ, Fisher AG, Murry EA, eds. *Sensory Integration: Theory and Practice*. Philadelphia: FA Davis, 3–24.

Franklin L, Deitz J, Jirikowic T, Astley S (2008) Children with fetal alcohol spectrum disorder: problem behaviors and sensory processing. *Am J Occup Ther* **62**(3): 265–273.

Hanson E, Kalish LA, Bunce E, et al (2007) Use of complementary and alternative medicine among children diagnosed with autism spectrum disorder. *J Autism Dev Disord* **37**(4): 628–636.

Miller LJ, Reisman JE, McIntosh DN, Simon J (2001) An ecological model of sensory modulation: performance of children with fragile X syndrome, autism, attention-deficit/hyperactivity disorder, and sensory modulation dysfunction. In: Roley SS, Blanche EI, Schaarf RC, eds. *Understanding the Nature of Sensory Integration with Diverse Populations.* San Antonio, TX: Therapy Skill Builders, 57–88.

Mulligan S (2003a) Examining the evidence for occupational therapy using a sensory integration framework with children: part one. *Sensory Integration Special Interest Section Quarterly* **26**(1): 1–4.

Mulligan S (2003b) Examining the evidence for occupational therapy using a sensory integration framework with children: part two. *Sensory Integration Special Interest Section Quarterly* **26**(2): 1–5.

Ottenbacher KJ (2002) Research in sensory integration: empirical perceptions and progress. In: Bundy AC, et al, eds. *Sensory Integration: Theory and Practice.* Philadelphia: FA Davis, 387–397.

Parham LD, Mailloux Z (2001) Sensory integration. In: Case Smith J, ed. *Occupational Therapy for Children.* 4th edn. St Louis, MO: Mosby, 329–381.

Parham LD, Cohn ES, Spitzer S, et al (2007) Fidelity in sensory integration intervention research. *Am J Occup Ther* **61**: 216–227.

Pfeiffer B, Kinnealey M (2008) Autistic mannerisms reduced by sensory treatment. *ScienceDaily* 27 April 2008. <http://www.sciencedaily.com/releases/2008/04/080425102403.htm>.

Schaaf RC, Miller LJ (2005) Occupational therapy using a sensory integrative approach for children with developmental disabilities. *Ment Retard Dev Disabil Res Rev* **11**(2): 143–148. doi:10.1002/mrdd.20067.

Schneider ML, Moore CF, Gajewski L, et al (2007) Sensory processing disorder in a nonhuman primate model: Evidence for occupational therapy practice. *Am J Occup Ther* **61**: 247–253.

Schneider ML, Moore CF, DeJesus OT, Converse AK (2008) Prenatal stress influences on neurobehavior, stress reactivity, and dopaminergic function in rhesus macaques. In: Burbacher T, Sachett GP, Grant KS, eds. *Primate Models of Children's Health and Developmental Disabilities.* New York: Elsevier, Inc., 213–258.

Smith Roley S, Blanche EI, Schaaf RC (2001) *Understanding the Nature of Sensory Integration with Diverse Populations*. San Antonio, TX, USA: Therapy Skill Builders.

Uyanik M, Bumin G, Kayihan H (2003) Comparison of different therapy approaches in children with Down syndrome. *Pediatr Int* **45**(1): 68–73. doi:10.1046/j.1442-200X.2003.01670.x.

Vargas S, Camilli G (1999) A meta-analysis of research on sensory integration treatment. *Am J Occup Ther* **53**(2): 189–198.