Ayres Sensory Integration Therapy for a Child With Rett Syndrome: A Case Report

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ABSTRACT: Rett syndrome (RTT) is a neurodevelopmental disorder characterized by severe dyspraxia, hand stereotypies, and sensory processing issues for which there is no known treatment. This case describes a child with classic RTT and the child's responses to an Ayres Sensory Integration (ASI) treatment intervention (36 one-hour sessions, 3 per week). We coded and analyzed 36 detailed treatment notes to answer the following questions: What strategies and factors facilitated or interfered with participation in the intervention? What critical elements of treatment documentation might detect small changes in praxis and participation? How do patterns of motor or praxis milestones that emerge over time relate to this child's level of participation? We observed an increase in participation when the therapist incorporated elements of neurodevelopmental treatment (NDT) and motor learning theory– treatment strategies commonly used with children who have neuromotor conditions. This increase in participation in the ASI intervention emerged at approximately the same time that the therapist documented acquisition of new motor and praxis skills. We observed the importance of using: lateral movement activities to develop weight-shifting and bilateral coordination, rotary play to increase trunk rotation and improve postural transitions, and rhythm to promote continuing or initiating actions. The documentation of the specific amounts of assistance and prompting needed during treatment sessions was an important tool for tracking small yet meaningful responses to treatment. This case illustrates a novel use of ASI intervention supplemented with strategies that developed foundational skills, and the emergence of praxis and participation in the therapeutic intervention. We suggest further research is needed to determine efficacy of ASI for other children with this rare disorder.

KEYWORDS: Dyspraxia, functional movement, motor development, occupational therapy, participation, sensory processing

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Introduction

Rett syndrome (RTT) is a disabling neurodevelopmental disorder that primarily affects females and occurs in about one in every 10 000 live female births—roughly 7000 per year. Most cases are linked to mutation of the *MECP2* gene. Children with RTT appear to develop typically during early infancy then they go through a period of regression around 6 to 18 months. This involves loss of previously acquired spoken language and gross motor and functional hand skills. They also develop dyspraxia of hand use and gait, sensory processing issues, and persistent hand stereotypies.[1-3] Although difficult to assess, cognition may range from low average to severely impaired.[4]

The severity of the dyspraxia[5,6] associated with RTT combined with loss of previously acquired motor and speech/language skills severely limit active participation in nearly all facets of their lives. They may not be able to complete basic tasks of grasping or manipulating objects to feed or dress themselves, participate in school activities, or play with toys or peers.[2] Most depend on their families or caregivers for their personal care and require long-term specialized education and therapies. The impact and stress on these families can be very significant.[7] There are no studies that demonstrate effective treatment for their severe dyspraxia.[8]

Serum Brain-Derived Neurotrophic Factor (BDNF)—a protein important for synaptic formation and neuronal activity that underlies balance and motor learning is reduced in those with RTT.[9] Studies with *Mecp2* animal models housed in enriched environments (EE) (cages with climbing toys, a running wheel, textured objects changed every few days) found they developed an increase in the number of synapses in the brain, an increase in cerebellar BDNF expression, improvements in motor coordination, and a reduction of anxiety related behavior, compared those housed in barren cages.[9-11] In a study of 12 girls with *MECP2* mutations, Downs et al[12] found that BDNF increased and motor skills improved following an intervention that combined EE and targeted motor-learning techniques.

There are parallels between the *environments* described in EE studies and Ayres Sensory Integration (ASI) treatment,[13] an intervention developed to treat children who have sensory processing disorders, typically used by itself.[9,14,15] ASI is provided by specially trained therapists in a sensory rich play environment—a space that contains adapted swings, therapy balls, scooter boards/ramps, rotating/bouncing/climbing equipment, and a variety of multi-sensory materials.[9,12,15,16] According to ASI theory active engagement in a rich sensory-motor play environment with a just-right challenge promotes adaptive responses and neuroplastic changes[13] that improve skill, function and participation. Recent research of children with autism spectrum disorders suggests that ASI therapy heightens sensory awareness of the body's position and movement and

improves sensory processing and praxis abilities, which can lead to more active participation in important childhood occupations, including self-help, play and school activities.[13,16-19] An earlier quantitative study used an interrupted time series design to investigate the efficacy of ASI in developing functional reaching and grasping in five children with RTT/Rett-related disorders.[20] That study provided preliminary data suggesting ASI had small positive effects on grasping objects. To better understand possible underlying mechanisms of this earlier study, we conducted a rigorous qualitative analysis of treatment notes of one child in that study to explore the relationship between observed praxis milestones and the child's response to ASI treatment. Specifically, we addressed the following questions:

1. What strategies and factors facilitated or interfered with participation in ASI intervention?
2. What critical elements of treatment documentation may be useful to detect small changes in praxis and participation?
3. How do patterns of motor or praxis milestones that emerge over time relate to the child's level of participation?

Case Representation

Participant information

In the earlier ASI study,[20] the Institutional Review Board gave approval to enroll 5 participants within the full range of RTT/Rett-related disorders. Written parental consent was obtained for participation and publication of study findings, including follow up case reporting. Of the 5 children with RTT/Rett-related disorders, only 2 had diagnoses of classic RTT, and of the two, one had never ambulated. Since approximately 60%-80% of children with RTT are ambulatory at some point in their childhood,[21] we chose to study the child who ambulated, in order to be most representative of other school age children with this condition.

This child is an 8-year old girl diagnosed with classic RTT ($Mecp2^{R168X}$). She has severe dyspraxia, hand stereotypies, poor hand grasp, gait abnormalities, difficulty *initiating* motor actions and postural transitions, and GI issues. She requires full assistance to participate in all activities of daily living.

Table 1 summarizes this child's abilities and areas of need at baseline, based on the following sources: occupational therapy (OT) evaluation, including sensory processing assessment; sensory processing surveys completed by her teacher and mother; OT treatment notes prior to the intervention; multidisciplinary team reports of present levels of performance from the Individualized Education Plan (IEP); and team quarterly reports on the student's progress toward IEP goals. The team included her school physical therapist, speech language/communication therapist (SLP), classroom OT (different from study intervention therapist), school psychologist, teacher of students with visual impairment, music therapist, classroom teacher, and parent. A summary of her OT evaluation (Supplemental A), goals of the ASI intervention (Supplemental B), and description of ASI intervention (Supplemental C) were prepared by the OT providing the ASI intervention.

Relevant past interventions. School records show that primary concerns from at least 4 years prior to the intervention involved this participant's mobility, communication, sensory processing, and self-regulation issues, as well as extremely limited hand use and participation in classroom activities, play, and self-care routines. This child attended a private school for children with substantial special needs, and received the following traditional therapeutic services at school:

- Physical therapy: provided individual and group therapy for strengthening, balance, and mobility—specifically, increasing walking distance without stopping, and stair climbing
- Occupational therapy: addressed sensory modulation, joint mobility, hand skills, self-feeding, and oral motor function through individual and group therapy
- Speech and language pathology: addressed communication through individual therapy using a total communication approach relying on eye gaze
- Music therapy: to increase language and participation.

Quarterly reports over this 4-year period prior to the study intervention revealed little to no measurable change in hand function, praxis, mobility or gravitational insecurity. Throughout this period, this child needed full assistance for all postural transitions, personal care, and for using classroom tools and materials, as described in her Occupational Profile (Table 1).

Treatment

Sensory integration is the ability to sort, organize and make use of the information simultaneously received by our senses (visual, auditory, tactile, proprioceptive, and vestibular). Sensory processing difficulties often lead to delays in motor development, praxis, self-regulation, learning, and social participation.[17,22] ASI intervention addresses the child's ability to organize and use this sensory information to respond in an adaptive and organized way.[23] ASI is implemented by therapists with specialized training, who adhere to specific guidelines set by the *Ayres Sensory Integration Fidelity Measure*© (*ASIFM)* .[16] ASI therapy (different from sensory diet or sensory regulation strategies) is conducted in a space that contains a variety of adapted swings, therapy balls, scooter boards, rotating/bouncing/climbing equipment, and multi-sensory materials. The therapist playfully supports the child through just-right sensory-motor challenges on this equipment, to facilitate adaptive responses, new skill development, and ultimately increased participation.[16]

Since ASI is typically used to treat children who can initiate play on movable therapy/playground equipment with relatively

Table 1. Participant's occupational profile.

PARTICIPANT REPORT	
Parent and school professionals' concerns related to engagement in occupations & barriers to participation	This child is an 8 y/o girl with classic Rett syndrome (RTT) who needs full assistance for postural transitions, has severe dyspraxia and hand stereotypies, poor sustained grasp and functional hand use, no spoken language, and limited active participation in classroom/school, self-help & play activities. She needs full physical assistance for holding/using classroom tools/toys/eating utensils, and for dressing, grooming, and toileting. She demonstrates high level of arousal/increased baseline body tension. She has significant gravitational insecurity walking, approaching steps/elevator, & during transitions—evidenced by distressed affect, grabbing helper, rigid posture, and/or freezing in place. She walks rigidly with irregular rhythm, stopping after every few steps. She strongly avoids trunk rotation when assisted to change body position. She needs full assistance to go from floor-sitting to standing and vice versa. She is unable to crawl or roll She relies on a total communication system based on eye-gaze. She is non-speaking. She has frequent GI discomfort. Often requires venting of her G-J tube.
Occupations in which child is successful	She uses her hands to manipulate soft gel toys—only texture she sustains She walks (dysrhythmic and stops after a few steps) She maintains sitting balance on chair, and on adapted swings when holding onto ropes and movement is gentle and predictable. She is very social and engages with others non-verbally with her quick smile and intense eye contact.
Occupational history	She lives w/ her mother and 2 siblings who lead active lives. She attends a publicly funded, private school for children with significant special needs.
Performance patterns	She requires moderate to full physical support to use her hands to participate in school, self-help & play activities, and to change body position. She expresses herself using facial expression and vocalization. GI discomfort, gravitational insecurity, dyspraxia, & hand stereotypies *hinder* occupational performance.
ENVIRONMENT AND CONTEXT	
Physical, Social, Cultural, Personal	Her transdisciplinary school team (PT, OT, SLP, 3/6 teacher/student ratio) *supports* engagement at school and her loving family carries over school recommendations at home. There are no barriers to engagement in her adapted school setting.
PARTICIPANT GOALS	
Participant and Team's priorities and desired outcomes	*Summary of targeted goals*: She will be able to transition and move thru her environment & on/off play equipment with less fear and assistance. She will initiate reaching to and sustaining grasp on objects (all textures), and use objects functionally to participate more actively in valued student and childhood occupations

See Supplemental B Sensory Processing Evaluation Summary.

minimal support or assistance, we anticipated that this child would require several accommodations to achieve fidelity to ASI and to maximize the functional outcome of ASI therapy. These included: the use of an individualized, total communication system to determine the child's activity choice; adaptions to the physical environment and therapy equipment to accommodate for difficulty initiating movement and getting on/off therapy equipment, and to promote safety; cueing to participate in postural transitions; and extended wait-time (30 seconds) after cues to promote initiation of intentional movement and participation.[24,25] Frequency and duration of the intervention were set at 36 sessions, 3 hours per week over a 3-month period based on review of ASI dosage literature.[26] Staff from The Koomar Center/Spiral Foundation affirmed the fidelity of the ASI intervention using the *ASIFM* .[16] This included a site visit (for space, equipment and safety checks) and remote scoring of some hour-long video recordings of treatment sessions to verify required process elements.

Method for analysis

This case report focuses on findings from one data source—treatment notes written after each intervention session by the treating OT. These notes were recorded on an Excel spreadsheet designed to document details of this participant's responses during treatment sessions and the OT's adherence to ASI fidelity.[16] On this spreadsheet the therapist recorded information pertaining to targeted goals, adaptive behaviors, quality of movement (eg, trunk rotation and weight-shifting), level of cues/assistance/participation, the child's activity choices and how they were determined, and factors that interfered with and/or supported the development of praxis, sensory processing, and participation.

We systematically examined treatment notes using a priori codes based on ASI theory and best practices in OT. We used 8 categorical a priori codes, with more finite codes (sub-codes) positioned within each of the 9 categorical codes.

Table 2 identifies the coding categories and sub-codes. We primarily applied descriptive codes (to label/name observations), process codes (to label observable actions), and magnitude codes (to label levels of performance) as described by Miles et al.[27] The a priori categorical codes were: (1) sensory processing; (2) postural functions; (3) bilateral integration; (4) visual processing; (5) praxis; (6) participation; (7) strategies; and (8) interfering factors. Refer to the Coding Manual (Supplemental D) for detailed descriptions of coding categories and sub-codes.

When coding and analyzing the data, we followed guidelines for establishing trustworthiness outlined by Braun and Clarke,[28] Nowell et al,[29] and Creswell.[30] These included peer debriefing, triangulation, and maintaining an audit trail of judgments and decisions. We describe the methods for documenting baseline functioning and measuring responses to treatment in our tables and supplemental files. Raw data that includes rich descriptions of the ASI intervention and participant responses during each intervention session is provided in Supplementals E and F. The authors' data analysis can be cross-checked with all supplementals and tables.

Two OTs, with Sensory Integration Certification and expertise with RTT and ASI, independently read through all notes to gain a holistic overview before conducting 4 systematic *passes* through the treatment notes (rounds of coding). Both therapists were study authors—one was the treating therapist during this intervention.

After separately (blind) coding each pass, these therapists compared codes, discussed differences, and came to a consensus on the appropriate codes and emerging themes. In the first pass, we analyzed treatment notes using a priori codes. During the second pass, we synthesized treatment notes into a chart noting areas of interest and themes for further examination (Supplemental E). This chart also identified strategies that supported as well as factors that interfered with engagement and function. In the third pass, we studied the relationship between levels of participation and the emergence of sensory motor/praxis milestones (Supplemental F). For this chart, we assigned scores from zero (none) to 5 (independent) representing levels of assistance and/or participation of tracked items over the 36 sessions, and color-coded treatment sessions to indicate when highest levels of function/participation were reached and reliably maintained.

We condensed this chart into a timeline of *weekly averages* of sensory and praxis milestones, to view week-to-week relationships and participation patterns (Table 3). In our fourth pass, we further analyzed temporal associations between emerging sensory processing, acquired motor milestones, and levels of participation.

Outcomes

During our coding process, we examined the following: factors that interfered with this child's participation in ASI sessions; strategies that supported access to and promoted participation; critical elements of documentation that captured subtle responses to treatment; and patterns of motor or praxis milestones that emerged and relationship to participation.

Table 4 outlines the factors that adversely affected this child's active participation in the ASI therapy process. The severity of her dyspraxia and hand stereotypies, and issues with neuro-motor delays, sensory processing, communication, and cognition necessitated varying levels of assistance/cues, particularly during the first month of treatment. The therapist assessed the child's tolerability of the treatment during each session. Medical issues occasionally affected her participation, including gastrointestinal discomfort, upper respiratory infections, and fever. During those events the therapist consulted with the school nurse, sometimes vented her g-tube, and/or re-scheduled the therapy session.

During our coding process, we identified several strategies the therapist used that were associated with increased participation in the ASI therapy sessions, over time (Table 5). These included elements of neurodevelopmental treatment (NDT)[31] and motor learning theory[32] that are often used by experienced therapists to treat individuals with neuromotor disorders.

A spreadsheet was designed to document fidelity to the ASI intervention and to capture subtle responses to treatment to help the therapist monitor progress and/or the need to modify the approach (see Table 6). The recording of specific cues and the least levels of prompting and assistance needed throughout treatment sessions, enabled us to detect small steps that signaled the emergence of new skills.[33]

Table 7 describes the interplay between emerging milestones and the child's active participation in ASI.

Discussion

Our in-depth analysis of treatment notes revealed important strategies that supported, as well as barriers that interfered with this child's participation in ASI therapy. We observed the importance of using lateral movement activities to develop weight-shifting and bilateral coordination, rotary play to increase trunk rotation and improve postural transitions, and rhythm to promote continuing or initiating actions. By recording specific cues and least levels of assistance needed throughout treatment sessions, we were able to detect small but significant gains that would not be picked up by standard scales or checklists.

Central to ASI is the child's active participation in all aspects of the intervention. In early sessions, this child's active involvement was very limited due to severe dyspraxia—that is, her inability to initiate purposeful movement. The therapist had to passively move her through many actions to help her sample activities to determine activity choice, transition on and

Table 2. Coding categories.

A PRIORI CATEGORICAL CODE	SUB-CODE	DESCRIPTION OR EXAMPLE
Sensory Processing (SP)		*(Proprioceptive, Vestibular, tactile, auditory, visual)*
Registration	SP-R	Orients to sensations
Discrimination	SP-D	Senses differences in sensory stimuli
Modulation	SP-M	Hyper-reactive, hypo-reactive, or typical response
Other	SP-O	Other sensory processing observations
Postural Functions (P)		
Postural stability	P-S	Maintains stable head/trunk control/postural orientation
Postural adjustments	P-A	Muscle activation to maintain balance or prepare to move
Postural transitions	P-T	Moving prone to supine to sit/quad/kneel/stand, vice versa
Other	P-O	Other postural observations
Bilateral Integration (BI)		
Crossing midline	B-C	Moving arm/leg across midline of body
Symmetrical bilateral	B-S	Each side of body doing same movement at same time
Reciprocal bilateral	B-R	Alternating movements between R & L sides of body
Asymmetrical bilateral	B-A	Each side of body doing a different movement or action
Other	B-O	eg,: Improved bilateral rhythm, etc.
Visual Processing (VP)		
Visual attention	VP-A	Attend to relevant visual info, look at task at hand
Joint attention	VP-JA	2 people attend to each other and to same object/event
Vis motor integration	VMI	Eyes and hands work together in coordinated way
Other	VP-O	eg,: Eyes crossed midline, vestibular-ocular reflex, etc.
Praxis (PX)		
Continues motions	PX-C	After practice motion faded or stopped, continues motion
Imitation	PX-IM	Copies/imitates therapist or other person in room
Ideation/motor plan	PX-I	Develops idea & executes motor action w/ no cues/assist
Other	PX-O	eg,: Actively approximates intended movement
Participation		*In transition/play/self-help/tx activities: trace to initiates*
Adaptive behaviors	PA	Rights trunk/balances/protectively extends/motor plans/reaches/grasps/differentiates 2 sides of body/crosses midline/visually attends, localizes or tracks to particip
Anecdotal observation	ANEC	Comments from team/parent on new participation in classroom/therapy/home/ community activity
Strategies (ST)		*(Adaptations to increase assess to ASI or participation)*
Equipm/tools adapted	ST-E	To address postural, bilateral, praxis or visual issues
Techniques adapted	ST-T	To address sensory, motor, and/or cognitive issues
Praxis adaptations	ST-P	eg,: Wait time after cues, practice motions/fading assist
Orthopedic adaptation	ST-OR	Due to decreased range of motion, scoliosis, etc.
Weakness adaptation	ST-W	eg,: Therapist eliminates gravity, partial assistance, etc.

(Continued)

Table 2. (Continued)

A PRIORI CATEGORICAL CODE	SUB-CODE	DESCRIPTION OR EXAMPLE
Other	ST-O	Other strategy to increase access
Interfering Factors (I)		*(Interrupted engagement/participation, part-all session)*
Fatigue/sleepy/asleep	I-F	Lethargy, low energy, and/or dozing off
Distracted	I-D	Attention diverted from activity at times
Agitated	I-A	Upset by internal or external sensations
Pain/sick/ill	I-PS	Discomfort from physical/medical issues
Stereotypies	I-S	Involuntary, repetitive, purposeless hand movements
Orthopedic challenges	I-OR	Joint contractures, scoliosis, stiffness, etc.
Weakness challenges	I-W	Decreased strength affects stability or active movement
Other	I-O	eg,: Discomfort, avoids prone, strict sensory preferences, gravitational insecurity, typical Rett symptoms such as poor praxis, involuntary release, stereotypies, etc.

Table 3. Sensory and praxis milestones: Weekly average of levels attained each week (3-sessions per week).

WEEKS OF INTERVENTION	1	2	3	4	5	6	7	8	9	10	11	12
Sensory Processing												
Hyper-responsive (H)	H	H	H	H	H	H	H	H	-	-	-	-
Minutes to moderate level of arousal	13	15	20	16	45	20	41	23	<1	3	6	14
Rotate trunk or lateral weight-shifted	0	1	1	1	3	**5***	5	5	5	5	5	5
Flexed trunk	0	1	1	2	3	**5***	4	5	5	5	5	5
Core Praxis, Bilateral Integration & Sequencing												
Chair-sit to stand	0	1	2	2	**3***	3	3	3	3	3	3	3
Floor-sit to quadruped	0	0	0	0	0	**5***	5	5	5	5	5	5
Floor-sit to quadruped to crawl	0	0	0	0	0	**4***	4	4	4	4	4	4
Floor-sit to kneel	0	0	1	1	1	1	3	**5***	5	5	5	5
Floor-sit to stand	0	0	1	1	1	2	1	**2***	2	2	2	3
Stand to kneel	0	0	1	2	2	2	1	**3***	2	3	2	4
Kneel to floor-sit	0	0	1	2	2	**4***	4	4	4	4	4	4
Stand to floor-sit	0	0	1	**2***	2	3	1	3	2	2	2	4
Moves swing with body/arms	0	0	1	1	3	3	1	3	**4***	4	4	4
Distal Praxis												
Reach toward object	0	0	1	3	1	2	**4***	4	4	4	4	3
Grasp non-gel object	0	0	0	0	0	0	1	**2***	2	3	2	3

Bold number with *= skill becomes predictably reliable at this level. Key: 0 = Full (100%) passive physical assistance required, no active participation. 1 = Moderate (75%) physical assistance is needed for her to continue part of a motion/action while assisted practice is faded. 2 = Minimal (25%) physical assist is needed to continue partial or entire motion/action as assisted practice is faded, OR sustained grasp once placed. 3 = Touch prompt and/or gesturing cue needed for her to initiate and complete at least part of a motion/action. 4 = Initiated after physically assisted practice was stopped, she resumed same action and completed at least part, OR initiated part of a sequence of an action w/o physical cues. 5 = Initiated and completed the action independently, or used this skill while performing another more complex functional action.

Table 4. Factors that interfered with participation during initial sessions of ASI intervention.

FACTOR	IMPACT ON FUNCTION OBSERVED AT THE OUTSET OF ASI INTERVENTION
Gross Motor: Poor weight-shifting and trunk rotation	Unable to transition self from floor sitting to standing, and vice versa Unable to crawl Walking not fluid, starts and stops every few steps Full assistance needed to get on/off therapy/playground equipment and to transition from floor sitting to standing and vice versa Poor balance reactions; body became rigid when balance was challenged The above contributed to gravitational/postural insecurity when moving on level and uneven surfaces
Sensory Processing	Gravitational insecurity contributed to: Hesitation or fear of getting on/off or playing on movable therapy/playground equipment, stepping onto elevator, going up/down stairs. Hyper-vigilance navigating busy/crowded hallway. Severe dyspraxia interfered with her ability to imitate or initiate new motor actions; Could not follow verbal or gesturing prompts
Fine motor: Hand stereotypies	Unable to sustain grasp on objects for more than one second, such as toys, school materials, clothing, food/eating utensils, etc. Reflexively releases Sometimes caused skin breakdown due to hand clenching and fingernails Needed full assistance to participate in self-help, school and play activities.
Communication and cognition	Non-speaking. Unable to follow verbal or gesturing prompts Required alternative means of communication to choose activities she desired and to ask to continue or stop activities, eg,: eye gaze at yes/no and picture icons, changes in affect, etc. Could not imitate Needed alternate means to teach her how to play on equipment and interact with toys/activities, eg,: taking arms/body through practice motions then pausing and waiting for her to replicate or continue the motions
Medical: Frequent upper respiratory infections and GI discomfort	Sometimes affected school attendance and/or full participation in some therapy activities Management of gastrointestinal discomfort (venting G-tube) sometimes interrupted or delayed treatment.

off equipment, and use her hands during play. The therapist applied motor learning theory by using tactile-proprioceptive input through passive practice motions to help the child feel the desired movements and develop a concept of the actions. This technique is commonly used by therapists in the treatment of children who have neuromotor dysfunction.[32,34] In this case, for example, the therapist provided multiple opportunities to passively practice component skills of trunk rotation and weight shifting during postural transitions and functional play activities, that led to the gradual emergence of active, independent rotation and weight shifting, and increasing participation. (See Supplemental E Treatment Notes—Columns 3-6)

These hypotheses are supported by the following research studies of motor learning involving neurotypical individuals. Chiyohara et al[35] showed that passive movement of the upper extremity through desired motions enhances proprioceptive acuity of those movements and improves efficiency and accuracy of motor production when assistance is removed. Bernardi et al[36] demonstrated that passive movements paired with positive reinforcement facilitated learning new motor actions and can be useful in early stages of motor learning. Rosenkranz and Rothwell[34] demonstrated that sensory attention activities without motor output influence motor learning. In theory, these techniques increase somatosensory awareness and facilitate emerging ideation of the task[5] and planning of new motor actions.

Our coding identified that the treating therapist incorporated elements of NDT and motor learning theory into many of the early ASI therapy sessions. These are tactile-proprioceptive techniques often used by experienced therapists to facilitate active movement for children with neuromotor involvement. At the outset of the intervention period, limited and rigid trunk movements interfered with this child's ability to participate in many of the activities integral to ASI. NDT handling techniques facilitated active weight-shifting and trunk rotation. Lateral and rotary movement activities on suspended/moving therapy equipment activated weight-shifting, trunk righting, and trunk rotation. These new core skills led to improved bilateral integration and active participation in postural transitions. As transitional movements became more automatic, she participated in ASI treatments with greater independence and confidence. Supplemental F demonstrates this progression.

This child's over-responsivity to movement/vestibular sensations initially contributed to avoidance of postural changes and certain types of play. This was measured by clinical observations of physical reactions when the child's balance was challenged such as when moving on therapy equipment– (Refer to *Sensory Processing and Modulation* column of the daily treatment note, Supplemental E). The therapist interpreted the following signs as over-responsivity: apprehensive facial expressions, increase in total body tension, distressed vocalization, strongly grasping

Table 5. Strategies that supported participation in ASI.

STRATEGY	DISCUSSION AND EXAMPLES
Elements of neurodevelopmental treatment techniques & motor learning theory to facilitate active movement	Incorporated neurodevelopmental treatment (NDT) handling techniques,[31] motor learning theory (physically assisted, practice motions),[32] and fading prompt hierarchy theory[33] Provided multiple opportunities to practice skills of trunk rotation and weight shifting during postural transitions and functional play activities throughout ASI sessions.
Rhythmic sensory input to facilitate continuation of movement	Rhythmicity of the therapist's voice, touch, movement cues, and visual stimuli helped the child anticipate and time the next step in motor sequences and more fluidly string together individual steps of motor tasks. Examples: When the therapist placed her hand on the child's shoulders and provided a light, rhythmic, side-to-side movement cue, she: (1) began walking more rhythmically with less pausing, (2) initiated stepping up onto stairs with less hesitancy/latency, compared to no cue (3) transitioned from floor-sitting to kneeling to standing with significantly less pausing. After upper extremity, rhythmic practice motions were paused, she resumed functional arm motion with less latency during some play activities.
Lateral movement to promote weight-shifting and bilateral coordination	Physically assisted lateral movement (on swings, therapy ball, etc.) facilitated righting reactions, coordination between opposite sides of her trunk, and timing of movements. As assistance was faded over time, automatic weight-shifting began to gradually emerge. She gradually began to use active trunk movements/weight-shifting to propel swings and to transition from floor-sitting to quadruped, high-kneeling and crawling. (See Supplemental F for timeline)
Rotary play to increase trunk rotation and trunk flexion	Following rotary play (alternating 180 degrees of turning on therapy swings/riding toys), the therapist documented modulation of tone in her trunk, increased excursion and ease of active trunk rotation during postural transitions, and emergence of functional asymmetrical movement and independent postural transitions. Graded vestibular/movement activities through many planes of 3-D space helped to decrease gravitational insecurity New adaptive responses/functional skills emerged once rotation was established, as in examples below: For the first time in session 16 and in all sessions thereafter she: Independently transitioned from floor-sitting to quadruped, (incorporating active trunk rotation) Independently flexed her trunk while transitioning from floor-sitting to quadruped For the first time in session 16, after physical cues were stopped, she: Transitioned from kneeling and quadruped to floor-sitting (and in nearly all subsequent sessions) She began crawling short distances toward a favorite toy, at times reciprocally (and in nearly all subsequent sessions). (See Supplemental F, blue highlighted cells)
Alternate communication strategies to interpret the child's activity choice	Because the child was unable to initiate play on her own and she was non-speaking, we used the following means to identify her activity choices: She walked to familiar, preferred equipment and made eye contact with the therapist to indicate her choice. After her therapist helped her sample various activities, the child demonstrated signs of pleasure or disinterest to indicate which she preferred. Therapist used yes/no cards, picture icons, eye gaze and changes in affect to communicate desire to start, continue, or stop activities After repeated exposure to preferred activities, she made activity choices more quickly, with less support.
Extra "Wait time" to promote ideation, motor planning and initiation of actions	We systematically waited at least 30 seconds (prescribed in her IEP) before repeating cues. This often resulted in her continuing or re-enacting motions to participate in activities for the first time. eg,: In some sessions, from #6 on, after assisted practice was faded/stopped, and given wait time and help sustaining grasp, she continued or re-enacted motions to: move a shoe toward her foot; move a marker to the paper and make marks; knock down a tower of blocks; move a ball to a target; re-start the swing she was sitting on; and to transition herself from standing to kneeling.
Identify and provide meaningful activities to increase participation	Finding meaningful play experiences, such as using favorite toys, enticed this child to put forth the effort needed to participate in somewhat challenging activities.

therapist or equipment, and avoidance of specific play activities. The therapist respected the child's body language and adjusted the activity accordingly. Over time, this child slowly accommodated to gradual increases in acceleration as well as movement in many different planes of 3-dimensional space while playing on suspension equipment. As behaviors associated with gravitational insecurity decreased, she began developing new adaptive responses and demonstrated greater enjoyment/participation in movement activities that she had initially avoided.[37,38]

Creating meaningful play experiences is central to ASI intervention. This is a challenge when treating children with significant communication issues and limited ability to interact with objects. While imaginative play can be very engaging for many children, this child was better motivated by concrete

Table 6. Critical elements of documentation to capture subtle responses to treatment.

DOCUMENTATION DESIGN	INFORMATION RECORDED
We created an Excel spreadsheet with a checklist to ensure ASI fidelity. We included a space for comments, to capture information not anticipated/not included in the checklist We provided a key* to record levels of assistance/cues/ participation	Therapy equipment and activities she chose/enjoyed/avoided, and how choices were determined Adaptations to equipment, space, and activities Quality of movement: eg, absence/presence/degree of trunk rotation & weight shifting, halting/smooth, automatic, etc. Adaptive responses and signs of praxis observed each session Sensory modulation observations (anxious/hyper-vigilant/relaxed, etc.) Response to treatment/progress on goals and objectives Factors that seemed to *interfere* with engagement, participation and performance Strategies that seemed to *support* engagement, performance, and participation. The type and amount of physical support, cues, and prompting needed to achieve her most active levels of participation during each session.

*Key: 0 = Full (100%) passive physical assistance required, no active participation. 1 = Moderate (75%) physical assistance is needed for her to continue part of a motion/action while assisted practice is faded. 2 = Minimal (25%) physical assist is needed to continue partial or entire motion/action as assisted practice is faded, OR sustained grasp once placed. 3 = Touch prompt and/or gesturing cue needed for her to initiate and complete at least part of a motion/action. 4 = Initiated: After physically assisted practice was stopped, she resumed same action and completed at least part, OR initiated part of a sequence of an action w/o physical cues. 5 = Initiated and completed the action independently, or used this skill to while performing another more complex functional action.

Table 7. Emerging motor/praxis milestones and relationship to participation.

GENERAL OBSERVATIONS	SPECIFIC OBSERVATIONS DOCUMENTED IN TREATMENT NOTES
When weight-shifting and trunk rotation became active and automatic, new functional motor skills began to emerge, followed by increased participation As bilateral integration improved and gravitational insecurity gradually decreased active participation in postural transitions emerged	Until session 8, she needed moderate to full physical assistance to shift her weight and rotate her trunk during postural transitions. Also, needed full assistance for floor-sit to stand & vice versa, and to crawl During session 15, she *initiated* trunk rotation without cues/assistance for 1st time during transitions, and maintained this core skill in all subsequent sessions. During session 16, she independently incorporated trunk flexion into her movements without cues for 1st time, and repeated in nearly all subsequent sessions During session 16, after being assisted to practice transitioning from floor-sitting to quadruped and vice versa (level 4 in Table 3 Key), she repeated the action and then began crawling for 1st time. She repeated transitioning and crawling at this level of cueing in all subsequent sessions. At the beginning of session 24, with her hand held tightly, she awkwardly walked through a ladder laid on the floor—stepping/tripping on most rungs, needing mod-max physical assist to weight shift and balance. Next she chose vigorous lateral swinging activities, challenging her balance, trunk righting, and weight shifting. At the end of this session, with hand *lightly* held and very slight weight shifting cues, she walked through the floor-ladder with much improved rhythmicity and accuracy, and almost no apprehension. She then walked back to her classroom with much less stopping and a more rhythmic gait. Beginning session 26, she used active trunk rotation and asymmetrical movements to get off riding toy (spin disk) low to the ground These progressions paralleled a gradual increase in participation in ASI sessions. See Supplementals E and F for other examples.
Active Participation in Postural Transitions Emerged as Gravitational Insecurity Gradually Decreased	During initial intervention sessions, she was tense while riding on nearly all gently moving therapy equipment, demonstrating an apprehensive demeanor, proximal/trunk rigidity, and distal fixing in her hands. By session 12, she showed improved comfort and tolerance for a variety of movement activities, and she began to respond to manual cues to weight-shift during some postural transitions. From session 14 on, she appeared more confident during postural transitions and played more actively on moving equipment. eg,: Sessions 14 to 17+: she attempted to propel therapy equipment using active trunk movements Session 16: she seemed relaxed & giggled during vigorous rotary play on swings. Session 16: she *initiated* floor-sit to quadruped transitions for the first time, and continued to do so independently from this session on. Session 20: for first time she *independently* transitioned from floor-sit to kneeling, and did this independently in nearly all subsequent sessions.

activities with highly preferred sensory attributes. Also, the therapist's playful use of self, and strategic placement of highly preferred materials and toys, became meaningful and strong incentives for her to attempt challenging activities involving postural transitions, crawling, reaching, and grasping. For example, placing a favorite gel-squishy toy at the opposite end of the mat enticed her to crawl for the first time. Likewise, dangling a favorite toy at arms-length enticed her to reach for

this target while riding on moving equipment. This led to reaching for objects with other less-preferred textures while both she and the object were moving, and suggests emerging ideation of a new motor task and projected action sequencing—both major sub-functions of praxis development.[39]

The therapist's use of rhythmicity through touch, voice/music/sound, and physical movement cues supported the child in 2 important ways. Rhythmic cues helped her anticipate the next step in motor sequences, such as when transitioning from floor-sit to standing. It also helped the child *time* her movements as the therapist faded or stopped physically assisted practice motions. Determining the timing of movement initiation is a critical process that precedes voluntary action.[40] Rhythmicity is an important element of praxis and integral to motor planning, bilateral motor coordination, and projected action sequences.[39]

The therapist waited at least 30 seconds for the child to respond to a cue before repeating it. "Wait time" after cues is recommended for children with RTT,[25] and 30 seconds is prescribed in this child's IEP. This strategy allowed her time to develop a concept of the task and to initiate a motor response.[5,41] This often resulted in the child continuing or re-enacting motions to participate in some activities for the first time.

Critical elements of documentation provided important insights into this child's response to treatment. Most existing assessment tools and checklists lack sensitivity to capture subtle changes that we might expect for a child with this complex condition. Table 3 outlines the key that the therapist used to record specific amounts of physical support and prompting needed to achieve optimal levels of participation.[33] This systematic recording revealed small changes over time in the child's quality of movement, sensory processing, praxis, adaptive responses, and participation. It also informed the therapist on effectiveness of treatment/techniques and/or the need for change. This documentation method proved to be highly effective for detecting the many small steps that signaled emergence of new skills.

More significant responses to treatment occurred in proximal praxis (functional trunk movements) compared to distal praxis (functional reach and grasping) during this 36-session intervention. This may be a function of the neuropathology of RTT—hand stereotypies and signature loss of hand function—or may be related to normal developmental progression from proximal to distal.[42] It poses the question of whether more significant arm/hand function would have eventually emerged if the intervention period had been longer.

Strengths and limitations

- The systematic and detailed recording of treatment notes from 36 hour-long sessions provided an in-depth description of how one child with RTT responded to ASI treatment. This analysis reveals the interplay between specific treatment strategies and the child's emerging skills and goal attainment.
- As with all analyses of individual cases, these observations are not generalizable to other children with RTT. Nevertheless, we believe that this rich qualitative data may generate important hypotheses for further research into this novel application of ASI for treating the severe dyspraxia associated with RTT.
- An important limitation to our analysis is social desirability bias—one of the authors provided all ASI treatments and treatment documentation. To minimize this risk, all treatment notes were coded independently by 2 study authors with expertise in RTT and ASI, with strict adherence to all measures outlined in our methods.

Conclusions

The previous ASI study measured the efficacy of ASI to develop functional reaching and grasping in 5 children with RTT/Rett-related disorders. In this case report, we conducted a thematic analysis of treatment notes from one of the children who had a diagnosis of classic RTT, to better understand underlying mechanisms of that earlier quantitative study. The severity of this child's dyspraxia, hand stereotypies, sensory processing issues, and cognitive and communication concerns created challenges, especially at the outset of the intervention. Our examination of these notes revealed strategies that promoted active participation in the ASI therapy process. It was through this coding that we discovered that elements of additional neuromotor treatment techniques contributed to this child's participation, through development of active weight shifting, trunk rotation, and bilateral integration. This report identifies patterns and possible relationships between a combination of therapeutic techniques/activities and this child's emerging praxis and active participation.

These observations and insights highlight the importance of careful, methodical recording of subtle responses to treatment so that therapists can adjust approaches and/or incorporate additional strategies, with the aim of maximizing treatment effectiveness. We discussed the importance of recording the hierarchy of prompting/cues that the child needs to produce adaptive responses. This systematic documentation of clinical observations captured subtle changes in praxis, function, and participation. We illustrated the novel use of ASI to treat severe dyspraxia of one child with RTT, with the hope that future research into its efficacy for this population will expand treatment options for others with this condition.

Declarations

Ethics approval and consent to participate

Ethical approval for the original study and follow up case reporting was granted by the Boston College Institutional Review Board (IRB Protocol #: 12.068.01). Procedures followed were in accordance with ethical standards. Authors confirm appropriate handling of confidentiality and data security. Parents signed formal written consents for participation in the

original study and publication of findings, including follow-up case reporting after attending an informational meeting conducted by principal investigators. All participation was voluntary.

Consent for publication

Written parental consent was obtained for participation and publication of study findings, including follow up case reporting.

Author contribution(s)

Karen Rocco: Conceptualization; Data curation; Formal analysis; Investigation; Project administration; Resources; Software; Supervision; Writing—original draft; Writing—review & editing. **Wendy Drobnyk:** Conceptualization; Data curation; Formal analysis; Investigation; Project administration; Resources; Supervision; Writing—original draft; Writing—review & editing. **Susan Bruce:** Conceptualization; Data curation; Formal analysis; Methodology; Supervision; Writing—original draft; Writing—review & editing. **Stephen B. Soumerai:** Formal analysis; Methodology; Supervision; Writing—review & editing.

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Competing interests

The author(s) declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Availability of data and materials

Data for this thematic analysis was derived from 36 hours of contemporaneous treatment notes. All pertinent treatment notes and derivative analyses are available within the narrative, tables and supplementals of this manuscript. Our original study on which this case report is based involved five children who attended one small school (<40 students) affiliated with Boston College. Given the small student body and sample size, we did not register our data in a repository in order to protect these individuals' rights to privacy.

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Supplemental material

Supplemental material for this article is available online.

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