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Case Study

A Case Study on Dandy Walker Syndrome: Early Intervention and Rehabilitation

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ABSTRACT

Dandy Walker Syndrome is hydrocephalus associated with a posterior fossa cyst and dysgenesis of the cerebellum.1 It is a rare abnormality of CNS with reported incidence of 1 in 25000 live births.2 This is a case of 5 year old male case with truncal attaxia, dysarthric, scanning speech, hypotonia, occular dysmetria, poor pursuit, poor fine motor control and poor balance and coordination. An early diagnosis resulted in early intervention in motor, speech, psychological and pre academics skills at child development centre. This case is successfully managed with conservative rehabilitation plan preparing him for normal school.

Keywords: *Hydrocephalus, Dysarthria, Truncal Attaxia, Rehabilitation*

Dandy Walker Syndrome is an autosomal dominant inherited disorder with incidence of 1 in 25-35000 live births.[2] It is a congenital brain malformation involving the cerebellum and the fluid filled spaces around it.[3] The key features are lack of coordination of axial musculature, psychomotor and growth retardation, hypotonia, strabismus, high arch palate and difficulty in motor learning.[4] It accounts for 1-4% of cases of antenatally detected hydrocephalus. About 45% cases are associated with chromosomal anomalies in gene focus 3q24[5]. Infants and older children can display symptoms differently. Diagnosis is normally made around 3-4 years of age but symptoms appear by age 1. The essential key is an early intervention. Infants may first display signs of delayed motor development and a progressive skull enlargement. They will be behind in skills like sitting, walking and talking.

CASE PRESENTATION

This is a case of 5 years old child who was admitted to child development centre in September 2015 with the chief complains of unable to stand, cannot walk without support, unable to speak age appropriately, throws objects on others and not going to school. Child was evaluated on following heading- History: Born at term was first issue to consanguineous

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parents and only affected case in the family. Personal history: the child is irritable and hyperactive. On physical examination child had abnormal motor development pattern (a w sitting and frog jumping), bilateral foot drop and poor hand manipulation and fine motor skills was reported. The child had cerebellar attaxia: where person is unable to sit on the bed without steadying themselves. There was no coordination of movement, decomposition of movement, dysmetria, past pointing and rebound. Romberg sign was positive. Eye examination showed ocular dysmetria and poor pursuit. Speech was dysarthic, scanning, having irregular emphasis on syllables. The child had many articulation errors which fall into three main categories

- Errors of differentiation and tuning
- Errors of coordination and sequencing
- Missing gestures

Behavior assessment observed that child was continuously licking his nose with tongue, restless, does not follow commands, hits other children and throws objects .child was totally dependent for activities of daily living like dressing, bathing and toilet skills. Child has shape, color concept.

Diagnostic Assessments: The child was assessed on following scale:

1. Portage guide to early intervention checklist: socialization skills 3 years, language skills 2 years, self help skills 3 years, motor 15 months, fine motor skills 2 years, and cognition skills 3 years.
2. Brief ataxia rating scale (BARS) consist of 5 subsets that evaluate walking capacity (8 pts), heel to shin test for decomposition (2-2, scored left and right), finger to nose test for decomposition and dysmetria (3-3 pts), dysarthria (3 pts), occular pursuit (2 pts). Child‘s total score was 23/30.
3. Gross motor fine motor scale (GMFM-88) consist of 5 subsets that evaluate lying (100%), sitting (100%), crawling and kneeling (78%),standing (7.6%) and walking (16.6%) of the child total score was 60.44%.
4. Receptive Expressive emergent language scale –Extended (REELS-EXTENDED) is used to identify receptive and expressive language problems and calculate receptive and expressive age of the child .the child’s RLA was: 24-27months and ELA was 20-22 mths. Bilateral hearing sensitivity was within normal limits.
5. On Denver Developmental Screening Test (DDST): The Developmental Age of the child was 1 year 11 months and Developmental Quotient was 50 indicating delay in developmental functioning.
6. On Vineland Social Maturity Scale (VSMS): The Child's Social Age was 1 year 8 months and Social Quotient was 43 indicating deficits in socio adaptive functioning.
7. On BASIC MR: The child was found significant in domains of temper tantrums.

INTERVENTIONS

After a detailed evaluation of the child short and long term goals were targeted and was given regular physiotherapy, speech therapy, cognitive training, behavior modification and pre academics training.

Physiotherapy: It focused on gross motor development: correction of abnormal pattern of motor development using verbal feedback. To improve pelvic stability reciprocal crawling was introduced.

Balance and coordination training: Non equilibrium exercises, star balance, Swiss ball for balance. For coordination: equilibrium exercises and frenkel exercises were used.
Fine motor development: constraint induced movement therapy was used to control his fine motor movements. Emphasis was given on proprioceptive neuromuscular facilitation techniques for training bimanual tasks.

Cognitive training: It focused on cognitive functioning: he was given problem solving training through tower of color rings, shape puzzle task and reasoning exercises.

Attention enhancement training: He was started with beading activities, and sorting images activity.

Memory training: With the help of brain toys like (wooden tree shape puzzle, colorful abacus beads) to improve his cognitive abilities with the size, color and number organization concept.

Behavior modification: He started screaming whenever he used to get excited. During sessions, he was instructed to make proper eye contact with verbal cues and good and bad behavior was explained to the child. Orientation of basic emotions and way to expressing the emotions was also teach him. He was told about few bad habits like licking nose, teeth grinding are not good which he does normally.

Psychosocial training: He was made to socialize with other play mates cooperatively. He was given instructions about to introduce himself first, when in group and show gratitude by saying "thank you' after the game with his play mates.

Speech therapy*:* Imitation technique, repetition technique, recitation technique was used. Respiratory exercises was also taught to the child. Blowing activities, exercise of tongue, lips and oral cavity and other techniques like counting aloud and biofeedback was also used.

Pre academics sessions*:* It included pre writing skills, math concept formation, fine motor development skills, reading skills and recitation of rhymes.

DISCUSSION

Every quarter the child performance was evaluated and new treatment goals were set. After 15 months the therapy gain of the child is as follows:

Physiotherapy:

The child has reduced w –sitting and frog jumping and now follows reciprocal crawling; child has improved standing balance and has started supported walking with short stepping gait. Child can walk 10 steps without any support. Fine motor control has improved example: improved timing of Pecs's board activity. Bimanual activities requiring coordination example: stringing beads has also improved. Child was reassessed on BARS and the score improved to 11/30 which infers the severity of ataxia has reduced significantly. On GMFM

scale, child motor skills improved and he scored 78.12%. On Portage scale, socializations skills improved to 4 years, language skills to 5 years, self help skills to 4 years, motor skills to 2 years, fine motor skills to 3 years and cognitive skills to 5 years.

Speech:
The child was reassessed on **REELS** scale. His **RLA** has improved from 24-27 month to 41/2 -5 years and ELA from 20-22 months to 3 ½ -4 years. For clarity of speech, repetition of words *ra, la, ta, na and* simple sentence recitation is done.

Behavior modification:
DDST - After assessments, the child **DA** was 2 years 7 months and **DQ** was 56.
VSMS - The child **SA** was 2 years 6 months and **SQ** was 54.
Basic MR Total score 4 suggesting no significant behavior problems

Pre academics:
Child had poor pencil grip and no reading /writing skills. Now the child can scribble standing line, slanting line and sleeping line. Child can write letter A & B. Child recognizes 10 colors. Child can read 3 letter words, read picture book of fruits/animals/birds /opposites/ vehicles/ part of body.

CONCLUSION

Dandy Walker Syndrome case is often misdiagnosed as athetoid cp and treated for the same. An early diagnosis, proper counseling to the parents and early rehabilitation can help child to overcome his disability. This case has been successfully managed with the conservative approach, which itself is a rare occurrence.

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Conflict of Interests: There are no conflict of interests.

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