

SPECIALIZED TRANSITION PROGRAM  
DISCHARGE EVALUATION  
NEUROPSYCHOLOGY

Date of Admission: 06/07/2010  
Date of Discharge: 08/27/2010  
Date Evaluation completed: 08/19/2010

**Reason for Referral:**

Haya is a 7 year-old little girl with history of resection of a right cerebellar pilocytic astrocytoma on September 10, 2005, with subsequent shunted hydrocephalus, seizure disorder, and global developmental delay. Haya was first seen at the Kennedy Krieger Institute (KKI) in July of 2008. She has returned at this time to the KKI Specialized Transition Program (STP) for intensive multidisciplinary evaluations and rehabilitation to monitor progress, maximize function, obtain updated equipment, and assist in outpatient treatment and educational planning. A neuropsychological/developmental evaluation was completed during this admission to determine profile of cognitive strengths and weaknesses.

**Evaluation Methods and Instruments:**

Mullen Scales of Early Learning (MSEL) – repeated  
record review, parent interview, observation

**History:**

Information regarding medical and developmental history was obtained from a review of available records, including the KKI neuropsychological report dated 7/18/08, and an interview with Haya's father. Haya lives in Kuwait City, Kuwait, with her parents, Mohammad and Maha, her 2 older sisters, ages 11 and 8, and her infant brother, age 8 months. Haya's siblings are all typically-developing. Haya also has a nanny who has been with her since birth. The family speaks primarily English to Haya at home. Family medical history is noncontributory to this case.

Haya was born at full term with average birthweight, after a pregnancy complicated by elevated sugar, which was treated successfully. Bilirubin was elevated, but resolved with treatment. Haya met developmental motor, language, and social milestones within normal limits, with first words by around 8 months and walking by 12 months. Haya's parents first became concerned about her development at around 7 months, when they noticed that her fingers were contracted. Soon after, she showed deterioration of her gait and loss of previous language skills. Haya's parents took her to a hospital in Kuwait, where genetic testing ruled out Rett syndrome, but no diagnosis was determined. Medical history is also notable for frequent ear infections as an infant, and removal of adenoids in May of 2005.

Due to continued deterioration, Haya's parents took her to the University of Freiburg Hospital in Germany in September of 2005, when she was 27 months old. An MRI of the brain on 9/8/05

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revealed a large cystic tumor of the right cerebellar hemisphere with severe compression and displacement of the brainstem, and obstructive hydrocephalus. The tumor was completely resected the following day, and was determined to be a Grade I astrocytoma. No radiation or chemotherapy was indicated. There has been no recurrence of the tumor. Haya initially appeared to recover well, then became somnolent. Repeated CT's of the brain showed bilateral subdural effusions, midline shift to the left, congestion of the lateral and third ventricles, and parenchymal lesions in right frontal and left occipital regions. Lumbar puncture indicated elevated intracranial pressure, which was treated emergently by drainage of cerebrospinal fluid. Following this intervention, Haya showed significant improvements in alertness and resumed talking, eating, and normal sleeping. She continued to exhibit right hemiplegia and ataxia. She was discharged home on 10/4/05, with plans for rehabilitation in Kuwait. A physical therapy note from January, 2006, reported that Haya had a palmar grasp and absence of upper extremity protective responses, and "...appears to be able to understand simple commands and is trying to verbalize..."

Haya returned to Germany, where a shunt was placed in the frontal horn of the right lateral ventricle on 1/12/06 for treatment of hydrocephalus. Speech/language therapy notes from May of 2006 indicated ongoing cognitive delay, with language skills at approximately the 6 to 12 month level. Haya was able to babble, laugh, and turn her head to avoid food, etc., but did not use gestures or consistently respond to her name, and "sometimes appears to be deaf."

A brain MRI in Kuwait in August of 2006 indicated that ventricles were still dilated. Haya returned to Germany, where follow-up evaluation on 8/14/06 determined that the shunt was functioning properly. Haya's severe developmental delays and chronically enlarged ventricles were attributed to protracted elevated intracranial pressure prior to her surgery in Sept., 2005, and were described as irreversible. Haya continued to receive outpatient physical and speech/language therapies in 2006, then attended the Hope School in Kuwait in 2007 and 2008. Hope is a school for children with special needs, which provides a structured education in a small-group setting with 2 teachers. Therapy notes over that interval indicated increased attention to sound and to toys, occasional gestures (wave bye-bye), attempts to imitate sounds, vocalizations to music, and attempts to crawl. However, notes indicated absence of pointing, inconsistent response to name, decreased visually-guided use of hands, poor postural control, and ongoing global developmental delay. A repeat MRI on 2/12/07 indicated no change in ventricle status.

On 1/12/08, Haya had a tonic-clonic seizure at home lasting approximately 15 minutes. She was taken to Mubarak Hospital, where a second seizure occurred, with apnea and cyanosis of the fingers. Seizures continued for approximately 1 week, then were successfully brought under control with phenytoin and Tegretol. Haya was admitted to KKI in July of 2008, where she had 2 additional seizures. An EEG during that admission indicated slowing, especially in the right temporo-frontal region, and focal epileptiform activity in the left temporal lobe, "...consistent with the presence of focal cerebral disturbance involving the entire right hemisphere, but

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sporadic spike waves over the left temporal lobe.” An audiology evaluation indicated normal hearing. Visual acuity was determined to be functional. Repeat MRI indicated a well-functioning shunt and no recurrence of tumor. A neuropsychological/developmental evaluation completed during the July, 2008 visit to KKI indicated significant cognitive and adaptive delays (MSEL Early Learning Composite standard score (SS) <49; Vineland II Adaptive Behavior Composite SS=45). At chronological age 61 months, age equivalencies on subscales of the Mullen clustered between 6 and 12 months.

Following her discharge from KKI and return home, Haya went back to Hope School on a part-time basis. She continued to have seizures approximately once a month. Medications were changed to Keppra and Trileptal. She had a 9-month interval with no seizures in 2009, then seizures recurred with increasing frequency beginning in January of 2010. Haya has been unable to attend school this year because of seizures and associated fatigue, as well as persistent sleep disruption. She has continued to receive OT and PT at home, but speech/language therapy has been discontinued due to lack of progress. Haya’s father describes 1 seizure in January of approximately 30 minutes duration with cyanosis, after which there may have been some deterioration in skills. No revisions of Haya’s ventroperitoneal (VP) shunt have been indicated since it was placed in 2006. As noted above, Haya returned to Baltimore with her family in the summer of 2010, and was admitted to KKI STP for intensive rehabilitation, updating of equipment, medical management of seizures, and monitoring of developmental status.

### **Behavioral Observations:**

Haya was observed numerous times over the course of her STP admission, both in individual evaluation sessions and in group activities. At her current chronological age of 7:3, she presents as an alert, generally calm little girl, of average size for age. She remains seated in an adapted stroller, or in an adapted chair with seatbelt for stability due to decreased trunk tone and control. Haya is incontinent. She is able to walk approximately 200 feet on indoor, even surfaces, with close supervision for ataxic gait. Haya eats a regular diet and thin liquids by mouth, and is making progress in self-feeding with an adapted spoon and a raking grasp, using either the right or the left hand with no clear dominance established. Eye contact is variable – sometimes fleeting and at other times intense and prolonged. Haya typically does not appear to visually inspect toys or other objects. She may touch them and rub them on her face. Haya frequently produces stereotypical finger movements with hands held up in front of her eyes (flapping, touching index fingers together). She occasionally grimaces, rocks with eyes closed, and has some writhing movements of the arms. There is some tongue-thrusting, which parents say is new over the past year. She is very difficult to engage. She does not consistently respond to her name, and often shows no reaction to loud noises nearby. She is often quiet, but also at times engages in extended vocalizations, consisting of sing-song like jargoning, with smiles and laughing. She responds in this way to music, and particularly to the familiar Barney songs, though laughing is sometimes spontaneous with no apparent precipitant. She appears to try to imitate sounds, but infrequently. She was not observed to use any words or meaningful gestures during our sessions.

She resists hand-over-hand activities, and may slouch in her chair or turn her head to avoid non-preferred activities. She is never aggressive, but will cry loudly when distressed. She sometimes leads adults to what she wants, but does not point. Attention span is very brief.

### **Validity:**

Instruments used are valid for this child and for the referral question(s). Instruments were selected which minimized demands for receptive or expressive language or fine-motor control. Items were untimed. Instructions were demonstrated and modeled whenever possible. If Haya's response could not be determined with certainty, items were repeated as many times as necessary to clarify. The evaluation was completed over the course of multiple 30-minute sessions, to maximize attention. Results of this evaluation are judged to be a valid representation of current cognitive and behavioral function.

### **Evaluation Results:**

#### **A. General Developmental Level**

In order to assess level of development across a number of domains, Haya was given the Mullen Scales of Early Learning (MSEL). The Mullen is an individually-administered comprehensive measure of cognitive functioning for infants and preschool children from birth through 68 months of age. The Mullen consists of approximately 150 structured activities, which cluster into 1 gross motor and 4 cognitive subscales as follows: Gross Motor, Visual Reception, Fine Motor, Receptive Language, and Expressive Language. For children such as Haya, who are outside of the age range for which normative data are available, raw scores may be converted into age equivalencies (AE). The AE represents the age within the normative group at which the obtained raw score falls within the average range. The four cognitive subscales of the Mullen were administered to Haya. Overall, AE's range from 5 to 13 months, indicating severe intellectual disability/delay. Scores in 3 of the 4 areas either stayed the same or showed small but measurable gains since the most recent previous testing in 2008.

**1). Visual Perceptual skills:** Items on this subscale assess visual-spatial awareness and processing of visual patterns, primarily through discrimination and memory tasks. This was the area of greatest delay for Haya, with her AE of 5 months at this time falling slightly below her AE of 7 months seen previously. As noted above, her visual acuity is thought to be within normal limits, but her functional use of vision is very atypical. She does not consistently fixate on and track even very salient stimuli, such as a bright reflective surface or face schematic. On the other hand, she sometimes demonstrates visual behaviors which are developmentally more advanced than tracking, such as searching for an item dropped off the side of her table. Haya does not typically appear to use vision to investigate her environment or to learn about unfamiliar objects. She does not typically attend to information presented to the visual modality only, particularly information presented at a distance. She is more likely to attend to items that

are moving. Her AE on the Visual Reception subscale falls at 5 months at this time, with isolated skills extending to the 12-month level.

**2). Fine Motor skills:** Items on this subscale emphasize the output aspect of visual perception. Though visual discrimination and memory are involved, tasks primarily measure fine-motor control with the dominant (or preferred) hand. Haya was able to complete items on this subscale at the 12 month level. She was able, for example, to use 2 hands together at times and to turn pages in a cardboard book. She is not yet imitating a crayon line, putting pennies in a slot, or stacking blocks vertically. This score indicates a gain of 5 months since previous testing 2 years ago. There was minimal to no scatter of additional skills above the 12-month level.

**3). Receptive Language:** This subtest assesses comprehension of spoken information while minimizing the complexity of motor planning and spoken language demands. Haya's raw score of 14 on this subtest places her at an AE of 13 months at this time. This represents a gain of 6 months over the past 2 years. For example, she appears to recognize her name and understand the word "no" at times, and is able to follow a simple direction with and without gesture (such as "turn and sit"). She was not, however, able to indicate understanding of "wh.." questions, such as "Where is the door?" by looking, gesturing, or any other verbal or nonverbal response. She was not able to point to any named body parts on a picture of a doll, on the examiner, or on herself.

**4.) Expressive Language:** In terms of expressive language, Haya's skills lag slightly behind her language comprehension (receptive language), but the discrepancy is small. Her current AE of 12 months is the same as the score she obtained in 2008, which may suggest that she is not gaining skills in this area at the same rate as she is in the area of receptive language. At this time, Haya produces jargonizing sounds (i.e. unintelligible sounds but with the intonation and rhythm of conversational speech), and combines jargonizing with gestures. On both receptive and expressive language subtests, minimal scatter was noted beyond ceiling scores of 12-13 months.

Age equivalency scores on the 4 cognitive subtests of the Mullen are summarized in the table below. Age equivalencies from testing in 2008 are also included for comparison.

Mullen Subscale	Age Equivalency (months) – 2010 evaluation	Age Equivalency (months) – 2008 evaluation
Visual Reception	5	7
Fine Motor	12	7
Receptive language	13	6
Expressive Language	12	12

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**Summary/Impressions:** Haya is a 7 year-old little girl with history of complete resection of a grade I pilocytic astrocytoma of the right cerebellar hemisphere in September of 2005, obstructive hydrocephalus, now shunted but with persistent ventricle enlargement, seizure disorder, and global developmental delay, who was admitted to the KKI STP from June 7, 2010 through August 27, 2010, for intensive multidisciplinary rehabilitation and evaluations to maximize function, update equipment, update and modify treatment plans, describe current levels of function, and manage seizures. Results of the cognitive/developmental evaluation completed during this admission can be briefly summarized as follows:

- Haya has made a number of functional gains since her last admission to KKI in 2008, as well as over the course of this admission to STP. All of these gains are important, though not all are of the type sampled in standardized cognitive testing. Gains have been noted in walking, grasping, self-feeding, expressing preferences in food and activities, awareness of environment, frequency and variety of vocalizations, response to music, and expressing wants and needs through nonverbal methods.
- At this time (chronological age 87 months), standardized assessment with the 4 cognitive subscales of the Mullen indicates developmental level at the 5 to 13 month range. Severe global developmental delay is apparent, though use of the visual modality for exploration and problem-solving appears to be somewhat weaker than language-based function. The profile of scores indicates small but measurable gains of 5 to 7 months over the past 2 years since the previous evaluation. Though, as noted above, test scores can never fully describe an individual, the items sampled are known to be representative of the broader “streams” of development and to have predictive validity regarding outcome. Predictive validity increases with consistency of findings over time.
- Results of this evaluation are consistent with severe to profound persistent intellectual impairment (mental retardation). Haya’s condition appears to be stable and chronic, with no evidence of deterioration. There is also no evidence to counter the conclusions of the physicians in Germany that the brain injury and cognitive impairment are irreversible. It is likely that they are due to multiple factors, including the initial tumor, surgery, pre-and post surgery obstructive and malabsorptive hydrocephalus, ongoing seizures, and to a lesser degree, possible effects of seizure medication.
- If development continues at the current trajectory, Haya can be expected to plateau developmentally at the 3 to 4 year-old level. There is, however, no way to know for certain that the current trajectory will continue unaltered. It may change due to new developments in treatment, or due to unknown and unpredictable factors.

**Recommendations:** A number of excellent recommendations were contained in the KKI Neuropsychological report of 2008, and many remain appropriate at this time. We recommend review of that document by Haya's caregivers and her educational team. Additional recommendations include:

- Return to a full day of school in the previous setting (Hope) or a similar program for children with special needs.
- Haya would benefit from an educational program that could incorporate elements of the discrete trials and Applied Behavior Analysis techniques that have been successful with children with symptoms of autism. Some resources for parents and professional educators can be obtained from websites of the following organizations: Autism Speaks, Autism Society of America, Autism Internet Modules, Pyramid Educational Services, Mayer-Johnson products and services, and Division TEACCH at the University of North Carolina, USA.
- Return to regular OT, PT, and speech/language therapy, at school and/or on an outpatient basis. See the STP therapists' reports for treatment goals. Particular emphasis should be placed on maximizing Haya's use of the Super Talker assistive communication device trialed with success at STP. Train home caregivers in techniques for providing therapeutic interventions at home between formal therapy sessions. Staff at STP will be happy to consult by phone or email with therapists in Kuwait regarding Haya's care.
- Follow KKI Behavioral Psychologist's recommendations for treatment of disordered sleep cycles.
- Use the assistive communication devices (2 disc switches and Super Talker) at home. Pair picture icons on devices with real objects, to assist in teaching symbol use for communication. Gradually eliminate the real objects and expand the menu of icons/choices as Haya gains proficiency.
- Encourage self-feeding and expression of choices using communication devices during mealtimes.
- Incorporate multimodal techniques into therapies and interactions whenever possible. Music and other auditory stimulation is particularly advantageous for Haya. Other modalities, such as touch, proprioception, and smell, should also be incorporated into her daily activities.

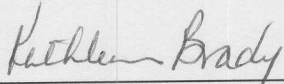
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- Provide access to a variety of toys appropriate for children at the 1 to 2 year-old developmental level. Toys demonstrating cause and effect relationships (for example, toys that play music and light up in response to a particular action) are especially valuable.
- We would like to see Haya again in approximately a year's time, to monitor progress and adjust recommendations.

I have enjoyed working with Haya and her family. Results of this evaluation were discussed with Haya's father prior to her discharge. If there are any additional questions about this report or if I can be of other assistance at any time, please feel free to contact me at (443) 923-4572, or at [bradyk@kennedykrieger.org](mailto:bradyk@kennedykrieger.org).



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*ICD-9: 225.0 Benign neoplasm of the brain, 345.8 epilepsy  
96118: 8 hrs.*