NORTHWEST REGIONAL PROGRAM
5825 NW Ray Circle
Hillsboro, Oregon 97124
503-690-5428

Functional Vision Assessment

Name: Matthew Wulf
D.O.B. 12/3/98
District: Hillsboro
Case Manager: Lorraine Duke-Atoui
Evaluator: Tambra Slack

Eval. Date: 6/30/98
Parents: Brian & Joyce Wulf
Address: 7295 NW Roy Rd.
Hillsboro, Oregon 97113
Phone: 648-6076

Purpose of Report: Matthew was referred to Vision Services to determine if he is eligible to receive services as a child with a vision impairment.

Eligibility Recommendation: Based on the following information, eligibility for vision services is recommended.

Background/Eye Report Information: Matthew is a 7-month-old child with Infantile Refsum's Disease. Refsum's Disease is a metabolic disorder, which results in decreased visual and auditory skills, developmental delay, and hypotonia to varying degrees. Matthew is under the care of a physician in Spain who is conducting research into a treatment for the disease, which includes the supplementation of DHA. Matthew has been receiving supplementation of DHA since his visit to Dr. Manuela Martinez in Barcelona Spain in May of 1998. Parents report that Matthew already appears to be improving.

A physician's statement dated may 27, 1998 by Dr. Manuela Martinez, M.D., which included vision information, was reviewed. The doctor reports that Matthew presented with "moderate axial hypotonia, psychomotor delay and visual deficit, although the infant could see light, bright colors, and fix on faces for a short time. Eye fundus examination did not find any alteration but the ERG was distinguished. Visual and brain stem evoked potentials were abnormal." Retinitis Pigmentosa is the eye condition associated with Refsum's Disease.

Assessment Tools:

Oregon Project for Children with Visual Impairments and Blindness
South Carolina Functional Vision Assessment
Columbia Regional Functional Vision Observation
Teller Visual Acuity Cards

Summary:

Matthew is aware of lights, colors and objects. He will fix on a face and track it using his head. Matthew will look directly at his mother's eyes and will watch her lip movements. He can track a bright colored toy horizontally and vertically using his head. Matthew watches his own hands and will often filter light through them. He reaches for his mother's face and hair and visually notices his surroundings. He has begun to visually

examine an object in his hand. It appears that he is beginning to look for a toy that has dropped out of his hand. Matthew appears to be aware of objects in all parts of his visual field.

Matthew had sluggish pupil responses to light suggesting a problem with light dark adaptations. Matthew is not yet tracking horizontally or in a circular pattern. He is not yet tracking vertically or horizontally without using his head. He had difficulty shifting gaze between two preferred objects, preferring to look at one briefly and then look away. Matthew appears to have difficulty changing focus from distant to near and from near to distant. Matthew tended to under reach for objects. Matthew is not yet smiling or playing with his image in a mirror. Because he appears to be most visually responsive and comfortable on his back, he has not had much opportunity to track moving toys or play on a tabletop.

Using the Teller Acuity Cards, Matthew's distance acuity appears, at this time, to be in the 20/200 range.

Recommendations:

1. It is recommended that Matthew receive services from a teacher of the visually impaired to assist parents in developing the use of his residual vision.
2. It is recommended that Matthew be given more opportunities to play with toys and observe his surrounding in an upright, seated position. This position would help develop tracking and scanning skills, and accommodation between near and distant points.
3. Reduce visual clutter and provide a high contrast to his play surface and objects presented to him.
4. It is recommended that Matthew be evaluated by an ophthalmologist.

Tambra Slack
Teacher of the Blind and Visually Impaired

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FUNCTIONAL VISION REPORT

Name: Matthew Wulf
D.O.B. 12-3-97
District: Hillsboro
Case Manager: Lorraine Duke-Atoui
Assessor: Ashley Talmadge

Assessment Dates: 10-14-98 & 10-22-98
Parents: Brian and Joyce Wulf
Address: 7295 NW Roy Road
Hillsboro, Oregon 97113
Phone: 648-6076

Purpose of Report

This assessment was conducted as a follow-up to a functional vision assessment conducted on 6-30-98. In the months following that assessment, Matthew's parents and his vision specialist observed many positive changes in his functional use of vision. This report provides updated information regarding Matthew's visual functioning.

Background and Ophthalmological Information

Matthew is an eleven-month-old boy diagnosed with infantile Refsum's disease, a peroxisomal disorder characterized by varying degrees of visual impairment, hearing impairment, developmental delay, and hypotonia. Retinitis pigmentosa, a progressive disorder of the retina resulting in the loss of peripheral vision, is associated with Refsum's disease. When Matthew was examined (3-30-98) at four months of age by Dr. Robert Steiner (Metabolic Clinic, Oregon Health Sciences University) he was found to be deficient in essential fatty acids including Docosahexaenoic acid (DHA) and arachidonic acid. Although there is no proven treatment for Refsum's disease, Dr. Steiner recommended that Matthew begin a regimen of DHA supplementation, given the apparent success of such treatment in several recent studies. Matthew has been included in a promising research study conducted by Dr. Manuela Martinez of the University Maternity Children's Hospital in Barcelona, Spain, and receives Docosahexaenoic acid ethylester (DHA-EE) supplementation as part of his treatment. He is monitored monthly by physicains at the Child Development and Rehabilitation Center of Oregon Health Sciences University. In his report of 6-9-98, Dr. Steiner details the many improvements noted by Matthew's treating physicians and parents (Brian and Joyce Wulf) since he began DHA treatment. Liver function was found to be improved, as was his tone. Joyce noted that he was much more content, babbling and entertaining himself for up to an hour; he was also sleeping through the night. Although his electroretinogram (ERG) was abnormal, a fundoscopic examination showed no evidence of the retinal changes associated with retinitis pigmentosa. Joyce reports that his most recent examination (11-4-98) also showed no evidence of retinitis pigmentosa. Matthew has a bilateral hearing loss (80%-90% loss in each ear). Joyce reports that his hearing aid improves his hearing

to a 30%-40% loss in each ear, sufficient to hear typical speech. At this time, he is sitting independently, is visually interested in his surroundings, and is attentive to sounds in his environment while wearing his hearing aid. He is not yet crawling, but is able to tolerate the four-point position for short periods of time with some assistance.

Assessment tools

1. The Oregon Project Skills Inventory for Visually Impaired and Blind Children ("Vision" section)
2. South Carolina Functional Vision Assessment ("Visual Skills Assessment section)
3. Teller Acuity Cards
4. LEA Heidi Fixation Sticks
5. Observation and parent interview

Results from the Functional Vision Assessment

Test environment: The assessment was conducted during two sessions in the living room at Matthew's home. Most often, Matthew was seated on the floor on a light green carpet. Furniture in the room included a pastel floral-patterned sofa, two plain-colored (mauve) armchairs, and several small tables and bookshelves covered with knick-knacks, books, and framed photographs. The room was illuminated by both natural lighting from a large picture window, and incandescent lighting from several lamps.

Vision Skills Checklist: Matthew has acquired 8 of the 13 skills listed in the "birth-1 year" category of the OR Project Skills Inventory (vision section). He stares at a source of light and visually explores his surroundings, and is usually able to maintain a visual focus on various objects for 5 or more seconds. He regards his own hands, is able to track objects across midline both horizontally and vertically (moving his head at the same time), and demonstrates visually-directed reach and grasp. Matthew does not yet track objects diagonally or in a circular movement. He often visually examines objects held in his hand before orally exploring the objects. Matthew rescues a toy dropped within his reach, and turns to look for objects out of view if there is an auditory component. Two more skills are emerging. Matthew is becoming more able to shift his gaze between two objects, and adding movement (shaking a toy, for example) is especially effective for gaining his attention in this regard. He will sometimes track a toy moving across a flat surface (a rolling ball, for instance), but he does not do this consistently. He was most successful at this skill when presented with a large object (an eight-inch blue textured ball, for instance) at very close range (within 12-14 inches). Matthew is not yet looking for toys that drop far from his reach (from a chair or car seat, for instance) unless the toy has an auditory component. He does not yet remove a cover to obtain a hidden toy, and does not respond to gestures (except smiling).

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Near acuity (formal): Results obtained using the Teller Acuity Cards would indicate that Matthew's visual acuity at near point (16 inches) is around 20/360 using both eyes together. This result might give the impression that his eyesight has worsened since the previous functional vision assessment. However, it must be noted that during this assessment Matthew quickly became quite disinterested in looking at the cards. Once his attention had been lost it was difficult to obtain responses, even to cards he had previously responded to quickly. Therefore these results cannot be considered reliable.

Near vision (informal): Results obtained by presenting red pompoms of various sizes at a distance of 12 inches indicate that Matthew's visual acuity at near point is more likely somewhere between 20/200 and 20/320. He gazed at 1/4-inch red pompoms presented on a white background, but did not appear to notice the 1/8-inch pompoms when presented at the same distance (12 inches). Thus, it appears that Matthew would be able to see an object of between 1/8- and 1/4-inch when presented at this distance.

Mid-range and distance vision (informal): Although it appears that Matthew is most visually aware of objects when presented within 18 inches (he was observed reaching for and grasping a variety of 3-to 6-inch toys when presented at this distance), he will respond visually to objects at greater distances when encouraged to do so. LEA Heidi fixation sticks (high contrast black-on-white faces on paddles of three sizes) were used to gauge the distance to which Matthew could maintain his visual focus on an object. He was able to maintain his focus on the 2-inch Heidi fixation stick as it was moved from a distance of 10 inches to approximately 2 feet. He maintained focus on the 4-inch Heidi face to about 39 inches, and on the 8-inch Heidi face to a little over 5 feet. In general, Matthew is not visually attentive to people and objects presented at distance, but his ability in this area appears to be improving. Joyce reported a recent incident in which Matthew was sitting in the living room, and Joyce was on the stairs about 10 feet from him. She called to him, and he visually located her. He was then reportedly able to visually follow her as she moved up the stairs. This occurred when the room was well illuminated with afternoon sunlight.

Visual fields: Informal visual field testing was performed using a penlight. As Matthew faced forward (his visual attention focused on a toy), a penlight was presented from his far right side and moved toward the center of his visual field at eye level about 14 inches from his head. This technique was used on the left side, and from above and below his central field. Matthew responded slowly at first to the light, and did not seem to notice it until it was approximately 45 degrees from center. (Typically, a child would look toward the light at about 75 degrees from center.) Matthew did respond more quickly after several trials (usually about 60 degrees from center), but often continued to respond more slowly when the light was presented on the left side. It must be noted that it was relatively difficult to draw Matthew's attention from one object to another on any occasion. As was previously noted, shifting gaze from one object to another is an emerging skill for Matthew. In order to perform this particular test, it is necessary to gain the child's attention centrally (with a toy or other visually interesting object). The child must then shift his attention off center when he sees the light. Thus, results from this visual field test must be regarded with caution; his peripheral fields may well be intact.

However, because of the possibility that he may develop retinitis pigmentosa (a condition which affects the peripheral vision), it will be important to continue to test his fields regularly. As he develops more sophisticated vision and communication skills, it will be easier to accurately assess his visual fields.

Ocular functions: Although Matthew did not demonstrate convergence during the assessment (the movement of the eyes toward the bridge of the nose as an object is moved toward the nose), his mother reports that he does demonstrate this ability in functional situations (when nursing, for example). Matthew's pupillary response (constriction) to a penlight was equal in each eye, but sluggish. As noted previously, Matthew was able to track objects presented within 14-16 inches both horizontally and vertically across midline. When tracking, he moves his head with his eyes. He is not yet tracking diagonally or in a circular motion.

Summary of Results

Matthew's functional visual acuity at near point appears to lie between 20/200 and 20/320. He is developing his ability to notice objects and people at distances beyond 3 feet. Pairing movement and auditory cues with the presentation of a visual stimulus helps to attract Matthew's visual attention. Several times during the assessment, Matthew did not appear to be aware of a toy/object until it was shaken and/or made a sound. Once his visual attention was obtained however, he engaged in much visual exploration of both his immediate environment (within a radius of 18 to 24 inches) and of toys he picked up. Testing of Matthew's visual fields could not be considered reliable because his ability to shift his visual attention from one item to another is just emerging. As his ability to shift focus develops, it will be important to re-test his visual fields. Matthew is an engaging little boy who remained happy, attentive, and playful for most of each testing session (about 40 minutes). His visual skills continue to develop and he is becoming more interested in visually exploring his surroundings at greater distances.

Recommendations

Ophthalmological:

1. It is important that Matthew continue to attend regular appointments with his local physicians, in which his ophthalmological issues are addressed. It will be important to monitor the condition of his eyes and to note any changes which may occur. It is recommended that Matthew be examined by an ophthalmologist.

   Questions for Matthew's parents to ask the eye doctor may include:

   a. How has Matthew's visual functioning changed?
   b. What has the most recent ERG shown? Has Matthew's retinal function remained stable?
   c. Have there been changes in Matthew's visual field? Do his visual fields appear to be intact?
   d. Is there anything we should do to prepare Matthew for his next visit?

2. Provide Matthew with a visor and/or sunglasses to protect his eyes from bright

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VISION UPDATE REPORT

Name: Matthew Wulf
Date of Report: 4-21-99
Vision Specialist: Ashley Talmadge
Case Manager: Lorraine Duke-Atoui

D.O.B. 12-3-97
Parents: Brian and Joyce Wulf
Address: 7295 Roy Road
Cornelius, OR 97113
District: Hillsboro

Purpose for Report

Matthew and his family are moving to North Carolina. This report provides an update on Matthew's progress in his acquisition and use of vision skills since his functional vision assessment was conducted ion October of 1998. It is hoped this information will be helpful in continuing an appropriate educational program designed to meet Matthew's unique needs as a child with a dual sensory impairment.

Background Information

Detailed information regarding Matthew's ophthalmological background is contained in a functional vision assessment report dated 10-22-98, and in medical reports prior to that date. Matthew subsequently had an eye examination and ERG on 11-4-98 with retinal specialist Dr. Richard Weleber (who has been following Matthew's case at Oregon Health Sciences University). The ERG revealed "severely subnormal responses of both rods and cones that were typical for disorders of peroxisomal biogenesis." Prior to dilation, Matthew's pupils were noted to be "definitely responsive to light but only weakly so." The optic nerve heads showed a "very slight peripapillary pallor." Foveal reflexes were not evident in either eye, although the macular region showed no substantial deterioration. Matthew's mother (Joyce Wulf) raised some questions regarding his 11-4-98 exam and ERG, and these questions are answered in a letter from Dr. Weleber in a letter dated 1-25-99. Dr. Weleber addressed issues related to the response time of the photoreceptors, loss of foveal responses, and optic atrophy. He also noted that the retinitis pigmentosa Matthew has (typical in cases of Infantile Refsum's Disease) does not result in the tunnel vision typically associated with RP. Rather, "the retinal dystrophy with IRD usually results in loss of central field first with retention of some peripheral field. The peripheral field is not entirely normal, and indeed, does become more affected as the individual gets older." Dr. Weleber noted that people with IRD probably use their peripheral field to view objects and people. Dr. Weleber recommended that Matthew be observed in regard to the direction of his gaze (does he look directly at people/objects, or does he appear to be viewing them from the side?), as this could provide clues as to what parts of his visual field he uses.

Update on Functional Use of Vision

Since Matthew's functional vision assessment of October 1998, he has acquired several more of the skills listed in the "Vision" section of the Oregon Project Skills Inventory. He is now much more consistently able to shift his visual attention between two objects, even when presented at different distances. He is also consistently able to track a toy as it moves across the floor, table, or tray in front of him, at distances of up to 4 feet. He inconsistently looks toward toys which fall to the floor while he is sitting in a chair or car seat. He will remove a cover from a partially-hidden toy, especially if the toy is highly motivating. Joyce reports that he is responding to some of her gestures and signs appropriately. Matthew is now able to track objects diagonally and in a circular motion. Matthew has increased his ability to maintain visual focus on objects and to visually inspect objects. He has become much more interested in finer detail. He has also increased his visual sphere, and typically interacts visually with objects and people within a 4-foot radius. When motivated, he will maintain focus on people and objects as they move into the 10-foot range. He is increasing his ability to use his vision while in motion, and is more apt to reach accurately for an object toward which he has been moving. (Previously, when moving toward an object he was likely to reach for the object before it was in grasping distance.) At this time, Matthew appears to be using his central vision to view objects and people; no head tilting or eye turns have been noted in this regard.

Progress on Specific Goals

Matthew is progressing well on his vision goals. Matthew often does take advantage of the best characteristics of toys; he does crumple paper, and the concept of rolling a ball or wheeled toy is emerging. (At this point, he is as likely to shake a small ball as to roll it.) Matthew is learning to place objects into containers, and will do this with lots of encouragement. He does show anticipation of certain events when given a sign (nursing, for instance); he is in the process of increasing the number of events for which he shows anticipation when given a visual cue. Matthew does search for partially hidden toys, if he is very motivated. For instance, he will remove a cover to obtain his favorite cup. He does not yet reach behind or move behind a barrier to obtain a toy. As stated before, Matthew is increasing his visual sphere and notices objects presented at greater distances. Although Matthew had been somewhat tactilely defensive when presented with unfamiliar textures, he is now able to tolerate a wider variety of textures. He voluntarily explores dry rice and other substances. Although he has not had a lot of experience with "messy" activities, he has tentatively explored some playdough. Matthew does visually inspect pictures in books, and sometimes will explore the pages tactilely. He does not yet operate sound strips or activate moveable parts in books.

It has been a pleasure to work with Matthew and his family, and to share in his progress. If there are any questions regarding this report, or Matthew's questions about my experiences with him over the last 6 months, please do not hesitate to contact me at 503-614-1634.

Ashley Talmadge

Ashley Talmadge
Teacher of the Visually Impaired