

Quality of Life: Issues in the Lives of Children and Families with MLD

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Objectives

- Definitions of Quality of Life

Three issues expressed by parents and children are:

- Discuss specific educational issues
- Discuss sibling-related issues
- Discuss psychosocial issues

For the meaning of life differs from man to man, from day to day and from hour to hour. What matters, therefore, is not the meaning of life in general but rather the specific meaning of a person's life at a given moment.

Viktor E. Frankl, M.D., Ph.D. (1905-1997) was an Austrian neurologist and psychiatrist as well as a Holocaust survivor.

What is Quality of Life?

- QoL is a multidimensional approach originated by the World Health Organization with 4 core domains: disease state and physical symptoms (physical dysfunction does not necessarily negatively impact overall adjustment), functional status (ability to perform a variety of age-appropriate daily activities), psychological functioning (in children/adults with MLD measuring psychological states independent of illness is problematic), and social functioning (ability to maintain social relations, and may be defined as the number and /or quality of social contacts).
- Other domains: "satisfaction with appearance," academic achievement, neuropsychological functioning, and overall satisfaction with life.
- HRQOL (Health related quality of life) refers to aspects of health and well-being influenced by disease and treatment in the health care system. It is not just the presence or absence of disease or infirmity.
- In addition to physician judgments, health status as perceived by the patient (or parent) provide an important perspective on the outcome of chronic disease and its treatment (patient satisfaction and perception of QoL).
- There is no universally accepted definition



Why measure QoL?

- To determine whether the benefits accrued to the child/adult after treatment sufficiently outweigh the risks.
- The benefit of survival in an otherwise inevitably fatal disease must be weighed against possible diminished QoL.
- Analysis of the factors contributing to benefits and risks must include the parent and child perception of whether a positive QoL has been attained.
- Study of the factors contributing to QoL will identify ways to improve it.

Educational Issues:



- Majority of children with MLD receive special education services through the Individuals with Disabilities Education Act (IDEA)...resulting in the parents as the advocate:
 - good special education is expensive and limited
 - parents often expend an enormous amount of energy and time attending meetings, writing letters, and negotiating for services to ensure the school provides
 - ~ Free appropriate public education that includes "... special education and related services designed to meet [the child's] unique needs and prepare them for further education, employment, and independent living..." 20 U.S.C. 1400 (d)

Symptoms (form) Drive the I.E.P.

Peripheral Nervous System (PNS) or Central Nervous System (CNS)

Late Infantile:

Loss of motor developmental milestones; problems walking; speech disturbances; difficulty feeding/swallowing; loss of memory; loss of vision; possible seizure

Juvenile:

Problems walking; loss of previously achieved physical and mental skills, such as developing incontinence; behavioral changes; muscle tremors, spasms, or loss of muscle tone; decreased attention span; possible seizures.

Adult/Teens:

Initial symptoms are change in personality and behavior leading to frequent misdiagnosis of psychiatric disorders; behavior that is increasingly impulsive or uninhibited; problems walking and stiffness in legs; numbness, tingling and pain in the hands and feet; progressive decline in intellectual abilities.

Educational Expertise

- Understanding the specific strengths/needs of child
- Educating school personnel
- Understanding how schools operate
- Special education law
- Definitions
- Tests and measurements
- Keeping records
- Writing good evidence letters
- Preparing for meetings



Resources for Educational Assistance

- www.PACER.org contains thousands of files for parents and children with disabilities
- For information and resources about education of children with disabilities, visit: NICHCY at: www.nichcy.org
- For websites for disabilities organizations and information groups, visit the Yellow Pages for Kids site at: www.yellowpagesforkids.com
- FindLaw is an encyclopedic law site. <http://www.findlaw.com>
- Wrightslaw at www.wrightslaw.com has a wealth of information about special education law and advocacy.
- If you need accurate information about children's disability and appropriate educational techniques visit: fetaweb.com, ncd.gov, and ldonline.org
- The National Protection and Advocacy site is at www.napas.org

Sample I.E.P items appropriate for a child with MLD

- Other Health Disability due to the diagnosis of metachromatic leukodystrophy (MLD).
- Maximum individualized services: speech and language therapy/augmented communication training and occupational/physical therapy in the school and/or the home setting; complexity of medical and educational needs, provided with an educational one-on-one paraprofessional individually and exclusively in the educational setting. The paraprofessional should meet the requirements that apply to the professional discipline in which she/he is providing for special education or related services and is consistent with any state-approved or state-recognized certification, licensing, registration, or other comparable qualifications; comprehensive and integrated program focus on the development of independent life skills.
- Although an important reason for placing a disabled student in a regular classroom is to maximize their academic achievement, this is not the only purpose. Many children with MLD benefit from their non-disabled classmates. These social experiences in a regular classroom cannot be duplicated in a special education class or separate school settings. At a minimum, your child should be integrated with typical peers into nonacademic activities such as lunch, recess, and physical education which should be adapted to meet their need for safety.

•Because the treatment for MLD may involve transplantation, many children qualify for home and hospital educational services to provide instruction to public school students who are unable to attend a regular school program due to a physical or emotional condition which is verified by a physician. Instructional services are available to all qualified students during convalescence or treatment time in a medical institution or therapeutic treatment center, or at the student's place of residence. The length of instruction for students in a full-day program is six hours per week; however, parents should know this time is not fixed and can be increased through a team decision.

•Individual Health-Care Plan (IHP), which includes an emergency health-care plan. The school nurse should be involved in initial and ongoing discussions developing the IHP, since the nurse will serve as the case manager who establishes the school treatment, emergency plans, coordinates the nursing care and educates the school staff in monitoring and treatment of symptoms. S/he has the responsibility for consulting and coordinating with the student's parents and health-care provider to establish a safe, therapeutic environment.

•Given the orthopedic/muscular issues children with MLD have, it is recommended they are evaluated by a physical and occupational therapist who is familiar with children with storage diseases and their specific issues. Also, because of muscle issues and the pain and discomfort involved, it is recommended the appropriate padding and sitting equipment be provided.

•Assistive technology should be evaluated to use on a daily basis that is used to increase, maintain, or improve functional capabilities. Assistive devices should be unrestricted for use and consistently/continually available during the day in order to maximize educational opportunities.

•It is important that those working in the school setting receive training in programming and utilization of a Alternative Communication (AAC) device in the school and home setting. Subsequently the child should learn how to operate the AAC that has been programmed to fit my child's needs in order to maintain and improve functional capabilities and to facilitate independence.

•Studies from the National Institutes of Child Health and Human Development have shown that for children with difficulties better learn through a multi-sensory teaching method which is the most effective teaching method. It is recommended that my child's educational program incorporate this teaching method. Instruction should not dwell on one curricular activity which promotes discouragement, a lack of motivation, and boredom. If my child does not learn a particular subject after repeated trials, such as spelling, than spelling should not be part of her goals and objectives.

•Due to the disease/MLD... will be exhibiting behavioral/emotional/psychological behaviors that do not fit the typical profile; therefore, the school psychologist (counselor) will need to be involved in my child's IEP. The Plan is as follows: ...call parents, a "go to person," how to remove from the classroom, what will calm/excite my child. A functional behavioral assessment (FBA) may be appropriate.

Sibling Related Issues:



Summary of research:

- Siblings of children with cancer are particularly vulnerable to depression, anger, anxiety, feelings of guilt, and social isolation. (Spinetta, 1981)
- Siblings have been identified as the most emotionally neglected and unhappy of all family members during serious childhood illnesses. (Chesler, 1987)
- Younger siblings were reported by parents to have more externalizing behavior problems (aggression, regression, school performance). (Ferrari, 1984)
- Older siblings are at increased risk for internalizing disorders (depression, anxiety, obsessive compulsive symptoms). (Stawski, 1996)
- One study showed that nearly one third of the siblings of bone marrow transplant patients showed anxiety, social withdrawal, externalizing symptoms, and moderate levels of post-traumatic stress disorder. (Packman, 1997)

Siblings

- Siblings are at risk for poor academic achievement, impaired social interactions, guilt, aggressiveness, withdrawal, anxiety, tearfulness, jealousy, anxiety. (Sahler, 1994 and 1997)
- There is increased sibling rivalry, anger, frustration, feelings of rejection, guilt, loneliness, sadness, confusion, and anxiety. (Kramer, 1981)
- Researchers noted emotional stress, sense of emotional deprivation, decrease in parental tolerance, anger, and guilt. (Murray, 1999)
- Nearly half (49%) of siblings reported mild post traumatic stress and 32% indicated moderate to severe levels. One fourth thought their sibling would die during treatment. (Alderfer, 2003)
- Good care for siblings includes making siblings feel useful and participative, providing useful, manageable information, and giving opportunities to share thoughts and feelings. Assistance includes emotional support, fair attention and family life. (Von Essen and Enskar, 2003)
- Siblings with more social support indicated significantly fewer symptoms of depression, anxiety, and fewer behavior problems than siblings with less social support. High level of social support plays a protective role in psychological adjustment of siblings of pediatric cancer patients. (Barrera, 2004)

What can we do...

- ❖ Encourage siblings to come to the hospital or doctor visits when possible so that:
 - * they can get to know the staff, who can reinforce that they are special too
 - * with patient and parental permission, staff can clarify medical information requested by the sibling
 - * the siblings can feel helpful by spending time with their brother or sister; however, do not make the sibling a care-giver with overwhelming responsibilities

- ❖ Seek the guidance of your hospital social worker, psychologist and child life specialist. They are available to help with all sibling issues

- ❖ Your community may have programs especially for siblings

Psychosocial Issues

Variables that contribute to psychosocial issues...

- Parental support system, SES, parenting ability, and parental stress/anxiety level and child's reaction to parental emotions
- Medical and CNS involvement: developmental level, rate of cognitive decline
- "Hassle factor" in terms of insurance companies, therapies, acquiring adaptive equipment, environmental changes to the home, family/relative issues, discipline of child with MLD, and financial burden

Synthesis of the research results: the needs of parents...

- ✗ The need for normality and certainty
- ✗ The need for information
- ✗ The need for partnership

As a Team...

- Help negotiate the health care system and ensure that patients' needs are met
- Work in legislative and organizational areas
- Act as advocates in managed-care organizations, health-related corporations and government agencies
- Work collaboratively with other health care providers, administrators of health care facilities, community agencies and government officials
- As educators and health information specialists, advocates work to empower others
- Protect and enhance patients' rights, and often act as agents of change in the health care system

Current and Future Quality of Life Studies

- Determine factors that influenced QoL status at baseline or follow-up including treatment
- Correlate these QoL outcomes with both parent and child perceived quality-of-life measures
- Correlate these QoL outcomes with biological markers such as enzyme levels, MRI findings
- Develop risk/benefit analyses as our sample size increases
- Disease specific of QoL at University of Minnesota
- Sibling study at University of Minnesota

The Team



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