Childhood Lupus

Lupus is a disease in which your own immune system attacks your own body. The immune system is the part of the body that normally fights against infections. In Lupus, the immune system is over-active. This causes inflammation in the:

- Skin.
- Joints.
- Blood vessels.
- Internal organs (kidneys, heart, lungs and brain).

Caring for a child with lupus is one of the greatest challenges a family can face. After the diagnosis is made, the first step for every parent is to learn as much as possible about the disease and the special needs of a child with lupus. Parents need to educate the child with lupus and their other children. It is also important to educate their child's teachers, family, and friends.

SYMPTOMS

At one time lupus was thought to be more severe in children than in adults. Most physicians no longer believe this. Children diagnosed with lupus often have been ill for a long time before the diagnosis is made. Initial symptoms are often vague and children are thought to have an infection or cancer before the diagnosis of lupus is made. At diagnosis, children have advanced disease and are more likely to have significant internal organ involvement than most adults with lupus. This is because many children are not recognized as having early lupus until the disease has worsened. Children with lupus often need to begin therapy soon after diagnosis.

DIAGNOSIS

The first step for every family is to be sure the diagnosis is correct. Lupus is a highly variable disease.

- Some children have obvious disease symptoms with:
  - Fever.
  - Rash.
  - Kidney involvement.
- Others may complain of:
  - Not feeling well.
  - Being tired or achy.
- Some children may look fine, but may have:
  - Blood in their urine.
  - Other unseen problems.
- A positive ANA (Antinuclear Antibody) is usually required to make the diagnosis. However, there are many children with positive ANA's who do not have lupus.

- Most caregivers rely on the American College of Rheumatology criteria for the diagnosis of lupus. It consists of 11 criteria. At least four criteria are needed to make a definitive diagnosis of lupus. Some children will be diagnosed with lupus even if they do not meet full criteria. In children, it is important to remember that more problems may develop over time. If you have questions about your child's diagnosis, contact your local chapter of the Lupus Foundation of America (LFA) for a list of experienced doctors in your area. The American College of Rheumatology Criteria are:
  - Malar rash – Redness over the cheeks and nose in a “butterfly” distribution.
  - Discoid Rash – raised, red patches that look like quarters.
  - Photosensitivity – A rash brought on by sun exposure.
  - Oral or nasal ulcers – Sores in the mouth and/or nose.
  - Arthritis – Swelling or pain in the joints.
  - Serositis – Fluid/Inflammation around the heart or lungs.

- Renal disease – Protein/blood in the urine or abnormal biopsy of the kidney.
- Neurological – Seizures and or psychosis not caused by any medicine.
- Hematologic – Hemolytic anemia (decreased red blood cells due to breakdown), leucopenia (decreased white blood cells), or thrombocytopenia (decreased platelets).
- ANA antibody - positive test.
- Tests of the immune system (immunologic) – a positive dsDNA test, a positive anti-Sm test, positive antiphospholipid antibody or a false positive syphilis test.

- Once the diagnosis is made, parents are immediately concerned about what will happen to their child. While lupus can be a severe and life-threatening disease, many children with lupus will do very well. The prognosis (what is likely to happen) of lupus in childhood depends on the severity of the internal organ involvement. Children with significant kidney or other internal organ disease require aggressive treatment. Mild rashes and arthritis may be easily controlled. However, lupus is unpredictable. It is difficult to know with certainty what the long-term outcome for a specific child will be. Lupus is a disease that has:
  - Remissions (no symptoms).
  - Flares (active disease or symptoms).

FOLLOWING YOUR CAREGIVER'S ORDERS

The severity of a child's lupus is determined by many factors that are beyond your control. However, some very important aspects of treatment are within your control. Paying attention to the doctors' instructions can make a major difference in the outcome of this disease. This includes:

- Taking medicine as directed.
- Having blood or urine tests when requested.
- Keeping scheduled appointments.

Children have special needs. It is important that those caring for the child (parents and medical professionals) understand these needs. Parents need to understand lupus. Parents need to:

- Know the reasons for the prescribed medicine.
- Know what problems to **promptly** report to the child's caregivers.

If you have questions, be sure to get answers. A good working relationship with the child's caregiver is one of the most important factors in the successful treatment of this disease. Everybody with lupus is different. However, much of the information and advice for children with lupus is the same as given to adults. You are encouraged to read the pamphlets available from the Lupus Foundation of America. These instructions emphasize the aspects of lupus and its treatment of children and young adults.

TREATMENT

Treatment for childhood lupus depends on the severity of the disease. The most important subject for parents of children with lupus is dealing with the side effects of the drugs.

Some of the lupus medications are:

- *Corticosteroids* (prednisone) and cytotoxic immunosuppressants can have unwanted side effects. The most obvious include: weight gain and a rounded face (*Cushingoid*) appearance, acne, unwanted hair growth, and stretch marks are the most obvious. These changes are easily visible and may upset you and your child. Over time, medications contribute to weak bones, poor growth in height, and fatty deposits in blood vessels that can affect blood flow (*atherosclerosis*). These side effects can be lessened by keeping the dosage as low as possible while still controlling the disease.
- Cytotoxic immunosuppressive drugs are one alternative to high doses of corticosteroids. These drugs are important to the parents of children with lupus. When used properly, immunosuppressives allow a dramatic reduction in the corticosteroid dosage. This is a major benefit to children. It limits the medical side effects of the corticosteroids and helps to keep children from getting obvious steroid side effects. However, cytotoxic drugs are not free of risks:
  - Short-term, they carry the risk of damage to the bone marrow that can cause bleeding. It can also make

the child unable to fight off an infection due to a weakened immune system. Careful monitoring minimizes these risks.

- Cytotoxic drugs may also may increase a child's risk of developing certain forms of cancer in the future.
- There is also evidence that immunosuppressive drugs may interfere with a child's ability to have children in the future. Caregivers must balance the risk of these future problems against the benefits of better control of the lupus and less corticosteroid use. Although these risks scare every parent, many children do extremely well with their lupus. Serious problems from these medications are rare.

ALTERNATIVE TREATMENTS

Parents may turn to non-conventional treatments such as herbal “remedies” or “miracle cures.” These alternative methods of treatment have not been properly studied for their effects on the human body. They must be subjected to research studies before they are used. Everyone caring for lupus is committed to finding better alternatives to current therapies. If there was any proof that alternative medications were helpful for lupus, the Lupus Foundation of America would know and doctors would be quick to use them.

BEHAVIOR AND RESPONSIBILITY

One of the most difficult problems for every parent is how to let their children “grow up” without letting them take unnecessary risks. This is even harder for parents of children with lupus. The key concept must be “as normal as possible.” For every decision you face with your child (school trip permissions, friends, sleepovers, etc.), ask yourself if you would let them go if they did not have lupus. The only big exception is exposure to excessive sunlight. Excessive sunlight may make lupus worse. A child with lupus may not be able to go to the beach for the day or sit out in the sun by the pool. Children with lupus should always wear sunscreen when out in the sun. If you are unsure, ask your caregiver.

EDUCATING YOUR CHILD

The feeling of being overwhelmed can be very real for both parents and children. However, educating yourself and your child can help to lessen many of the worries and concerns. How to approach your child depends on your child's age. Of course no two children are the same, but these general guidelines can help.

- For children younger than age 10, long explanations are not very helpful. During this period children rely on their parents to “fix everything” and have little sense of responsibility. While your child should know that they have lupus, you do not need to spend a lot of time trying to explain it. They will need to understand that lupus is a disease and that it is not their fault. It is not something they caught from someone and that they cannot "give" lupus to anyone else.
- It is important that your child understands the need to take their prescribed medicine. For older children you can begin to explain the names of the medicines and the doses. For smaller children this is not helpful. Sometimes parents go to great lengths to explain every detail to their children as if they were small adults. Under the age of 10, this information is often misunderstood. Despite the best intentions, it may do more harm than good.
- For children 10 and older, it is important to begin explaining the nature of lupus. The transition from a small child who must simply do what they are told to a mature self-reliant adult may be difficult. It is important that children with lupus be allowed to assume an increasingly greater role in taking their medications and making choices about their care.

EDUCATING OTHERS

- You will rapidly become aware that you have to educate the school teachers and leaders of organized activities in which your child participates. You will have to explain:
  - Absences for doctors' visits.
  - Fatigue.
  - Changed appearance.
  - Restrictions in activities.
- Lupus support groups are a good way to help both the parents and the child meet others who have lupus as

well as their families, learn more about the disease, and explore ways to cope.

HOME CARE INSTRUCTIONS

- You must understand that your child will have days when they are upset about having lupus just as you are upset that they have it! They will be upset about the side effects of their medicine and upset about the limitations placed on them. The fact that it is nobody's fault does not make it better. You must treat your child just as you would if they were angry about anything else. Occasional problems are normal. If these feelings become prolonged, you may need to seek professional help. Remember, there is no single solution that will work for everyone.
- There is a whole pamphlet dedicated to depression in lupus provided by the Lupus Foundation of America. It is very important that parents recognize if their child is becoming depressed. Children who become depressed about having lupus may stop taking their medication without telling their parents. This can be disastrous. Watch the medicine bottles. Are they getting empty? Are refills needed as often as expected? Even if you are the one administering the medication everyday, stop sometimes to watch and make sure the pills are being swallowed and not spit out when you turn around. You might be complying with all of the caregiver's advice while your child is not. If you discover that your child has not been taking their medication, talk about it with them, and be sure to let your caregiver know.
- It is important to remember that most children hate being sick. Often this hatred will be expressed as hatred of the pills, hatred of the blood tests, hatred of the doctor's visits, and hatred of advice from parents. This can often be dealt with by reasoned discussion and making sure the child knows why they have to do unpleasant things. At some point in this age group, they need to understand that doing the wrong thing could make them very sick. Few children in the pre-teen and early teenage group realize their actions may have permanent consequences that if they do the wrong thing. They cannot simply “be forgiven and start over.” If you cannot work it out with your child, discuss the situation with the child's caregiver. Some children may require psychological intervention. If not dealt with, a child or young adult's resentment about having lupus may result in risky behavior that can be disastrous or result in death.
- Ask the caregiver for help if you are having trouble getting through to your child. Not only does having lupus cause psychological problems, but lupus itself may cause problems with thinking. Sometimes a child who is refusing their medicine may need more medicine. It is important to understand the psychological problems as well as the effects that lupus can have on *cognitive* (thinking) impairment. If you believe your child needs help, the child's caregiver can help you find an experienced psychologist in your community.
- Even children who appear to be doing well may have questions they are afraid to ask. Young people in their middle and late teenage years are beginning to think about their life as adults. Children with lupus will wonder if they are going to be able to get married, have kids, and make a living. The answer should always be that you expect them to lead productive lives. Since most children with lupus will live well into adulthood, it is important to make sure they understand the importance of doing all the things necessary to achieve their goals.

COULD ANYONE ELSE IN MY FAMILY DEVELOP LUPUS?

Family studies of children with lupus have shown an increased frequency of blood (*serologic*) abnormalities in the mothers of children with lupus. Because usually these are healthy mothers without evidence of disease, this was surprising. This finding strongly supports the hypothesis that lupus results from the combination of a genetic predisposition and an environmental exposure.

FOR MORE INFORMATION:

The Lupus Foundation of America
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