

March 30, 2009

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**Re: Emergency Management Protocols**

Dear Deb:

I want to share with the FOD support group three tragic situations in different states where we represented families of a child with an FOD that received improper care in an emergency setting. In two cases, mismanagement led to severe brain injury and permanent disability. In the third case, the child died. With proper medical care, all three kids very likely would have suffered no harm.

All of the parents involved are extremely dedicated and well informed. All of the parents sought medical attention at the first sign of trouble. In two out of three cases, requests for glucose were ignored.

I have several observations that stem from litigating for these children and talking with metabolic experts around the country:

**Point 1: Most children with FODs should have a written emergency protocol.**

The medical standard of care now requires that a metabolic physician provide a written emergency treatment protocol for most children with an FOD. I have seen the consequences when a protocol is missing, and it is crucial for parents to press their physician to provide one.

As recently as last summer the parents of a one-year-old with VLCAD were not provided an emergency protocol. Their little girl was identified by newborn screening and followed by a metabolic physician at a large university hospital.

This girl's father took her to an emergency room one evening because of a viral illness, vomiting and poor feeding. The father told the ER staff that his daughter had VLCAD and needed glucose. Because the child's plasma glucose was above 70 and the child did not appear "critically ill," the doctor did not provide IV glucose; the child received IV normal saline instead. The whole time, incidentally, the parent believed that his daughter was getting IV glucose! Rather than recognizing the potential for rapid decompensation, the ED physicians discharged the child from the ER at around 1:00 a.m. Later that morning the child became unresponsive and died from cardiac arrest in the ambulance on the way back to the hospital.

This little girl had been admitted when ill twice before to the same hospital and had received glucose both times – but she had seen *different* physicians.

Parents and doctors have wide-ranging personalities and experience. Some doctors are more willing than others to listen to parents. Some parents have better skill communicating with doctors. A written emergency treatment protocol, signed by a current treating physician, levels the playing field and takes away these variables. It is a binding document in the medical world. **There are serious repercussions if a treating physician fails to follow a valid protocol without discussing any protocol deviation with the child's treating physician or another qualified physician.**

If your child has a disease which can result in rapid decompensation and does not have an emergency protocol letter, you should insist on one. It is helpful to have your protocol on file at the hospital, but it is also necessary to carry it. There is no guarantee that your local hospital will find it quickly or that your child will be near *that* hospital when ill. It is also necessary to have your metabolism specialist's cell phone or pager number, but because it is not always possible to reach your doctor immediately, a written protocol allows treatment to begin while waiting for the specialist to call back.

This protocol letter also should have a section addressed to EMTs. Some FOD kids decompensate very quickly and treatment by EMTs may be able to save a child. Early responders may be even more reluctant than emergency physicians to vary from their own protocols based solely on parental input. A written protocol will address whether EMTs should start an intraosseus line with glucose if they are unable to start an IV (a frequent occurrence in a small child). The protocol should address whether EMTs ought to administer glucagon, which is typically in every ALS protocol for a child thought to be hypoglycemic. Finally, the protocol should inform EMTs that your child can decompensate very quickly. They should be instructed to begin transport immediately, even if the child does not appear to them to be critically ill or if the Accu-Chek does not reveal profound hypoglycemia.

Importantly, a protocol should instruct caregivers to trust the parents and let healthcare providers know that you have been educated about your child's problem and know when your child is sick.

Protocols for sick management at school are not a substitute for a written physician/EMT emergency management protocol. If your child needs one type of protocol, he or she also should have the other.

**Point 2:      Make sure you have a protocol that is current and signed by your doctor.**

I am representing a girl in a lawsuit where a family went to an ER with an unsigned

protocol that was printed from the FOD Family Support Group website. They filled in data about their child and wrote the cell phone number of their metabolic physician on the front of the document. The ER doctor denies ever seeing the protocol or receiving the cell phone number.

I believe that the doctor has forgotten seeing the protocol because he didn't take it seriously. Certainly he did not follow the protocol. On admission, the child was in congestive heart failure but did not receive appropriate treatment for between 4 ½ to 5 hours. This beautiful young girl, who before that day had no disability whatsoever, suffered a cardiac arrest and brain injury and now must use a wheelchair.

Very likely the parents' pleas to follow the protocol and call their metabolic physician were ignored because they did not possess what the ER doctor felt was a "real protocol," signed by a metabolism specialist. Because the girl's glucose was over 70 and she did not appear to be "critically ill," the doctor waited hours until test results came back to contact the appropriate specialists and begin treatment. A current, signed protocol will help prevent difficult confrontations.

Some parents are concerned that a protocol will be insulting to doctors because it suggests that they "don't know what they are doing." The truth is that ER doctors know that patients with complex diseases often carry protocols. Far from being insulting, it is usually a relief to them.

**Point 3: Do not let the ER doctor fly solo.**

Parents of children with metabolic disorders have reported that ER doctors, on occasion, will not follow a protocol letter, particularly if the protocol is from an out-of-state physician.

In that situation, you must insist that the ER doctor speak with your child's metabolic physician. If the doctor will not call, then *you* must. It is very unlikely that an ER doctor will refuse to talk if you hand him/her a cell phone and say, "I have Dr. So-and-so on my phone. He/she is my child's treating metabolic doctor. He/she would like to speak to you immediately."

**If there is a problem reaching *your* metabolic physician and your child is critically ill, it is better to page a qualified metabolic doctor that you do *not* know – even at a metabolic center you have never visited – rather than letting an ER attending physician struggle alone treating a complex, life-threatening disease for which he/she has little experience.**

The main number at Children's Hospital in Philadelphia is 215-590-1000. You can call that number and ask for the metabolism fellow on call 24/7. The same is true of most large

academic centers connected to a children's hospital.

Another alternative, if you meet resistance to a protocol letter, is to ask for the hospital's administrator-on-call, or, if it's a larger hospital, there may actually be a "patient advocate." Those people are responsible for helping patients when there is a dispute over care. Such calls may be met initially with skepticism, but if you remain calm and point out that you have a valid, signed protocol that has been disregarded – it soon will be the doctor who has to answer questions, not you.

**Point 4: Always confirm that a glucose measurement was taken on your child and that the value has been communicated to the doctor.**

Another tragic outcome occurred because care providers forgot to do an Accu-Chek on a very sick baby in the ER. On presentation, blood was drawn for a basic metabolic panel (BMP), which includes a glucose level, but the lab did not report the glucose value for almost four hours. In the interim, the child, who turned out to have an FOD, suffered a severe brain injury from hypoglycemia.

The ER doctor assumed that the boy's initial glucose level was normal because she was not told it was abnormal: Nursing protocols at that hospital require both an Accu-Chek upon admission and a report to the attending physician if the glucose is abnormal. But in that busy ER on that day, the nurse simply forgot to do it. The attending doctor was not informed of a problem until the laboratory called the ER four hours later with the BMP results. The child's glucose on arrival had been 26! In the hospital, unfortunately, parents cannot assume anything. One can't assume that plasma glucose was measured or was promptly reported or that an IV connected to your child actually contains glucose.

Parents of FOD children should consider carrying "Insta-Glucose," which comes in three-packs for \$12.99 at Walgreens. This glucose gel can be absorbed in the mouth and is useful when there is a delay in providing glucose to your child for whatever reason.

**Point 5: Glucose sometimes is not the only answer.**

For many children with FODs (for example, MCAD), glucose is the critical treatment during decompensation. But many FODs are more complex. With VLCAD, a child who has been fasting may have a "normal" plasma glucose because the child is no longer able to utilize glucose on a cellular level. The plasma glucose is "normal" when the child is sick because the child can't use glucose – not because organs, like the heart, are getting enough glucose. In those situations, insulin (in addition to glucose) is required to shift the child's metabolic "thermostat" from breaking down fats to utilizing glucose.

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When faced with a critically ill child with VLCAD, doctors who are not familiar with FODs will have a hard time understanding why they should administer a glucose bolus and insulin to a child with a plasma glucose level of 115. A protocol should spell out at least the basics of glucose metabolism as it relates to FODs and urge a doctor to contact your metabolic physician immediately to discuss management.

### Conclusion

Parents and organizations like the FOD Family Support Group, OAA, and Save Babies Through Screening have changed the newborn screening landscape forever. The New York Times just reported that supplemental newborn screening for metabolic disorders is “now routine for newborns.” [<http://www.nytimes.com/2009/02/19/health/18screening.html?ref=health>] This is a far cry from what most of us remember just a few years ago. The blessing of early diagnosis will mean many more children will be treated pre-symptomatically; but that achievement will be hollow if kids do not get appropriate treatment in the ER. My experience with these situations reveals that a current, signed protocol is critical to assure optimum treatment when children with metabolic disorders are ill.

Sincerely,

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**[Please note – from Deb: This letter is also pertinent for our FOD adults, as well as children. There have been times our adults’ protocols have been totally dismissed despite their knowledge of their own disorder and its treatment, as written by their metabolic specialist.]**

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