“Little Miracle” Joshua’s Story

"Extra-Corporeal Membrane Oxygenation (ECMO) is a heart-lung bypass procedure, that allows the lungs and heart to "rest" and recover from conditions such as Meconium Aspiration and overwhelming infection (as in Joshua’s case below) as well as other pulmonary conditions such as pulmonary hypertension. In addition, it can be used as a "bridge" following open heart surgery, as it is more commonly used in the older pediatric patients (in the Pediatric Intensive Care Unit). It requires a team effort for successful implementation, including perfusionists, respiratory therapists, specially trained bedside nurses, as well as nurse practitioners and physicians. Joshua was typical of the patients that benefit from ECMO at Westchester Medical Center—he was literally on death's doorstep when he came for ECMO, and his parent's accounting truly reflects the concern that he would survive. Please read their gripping account of this life experience."

Lance Parton M.D.

"The word “Miracle” is often mentioned in our family household ever since our son Joshua Tyler was born on March 19, 2002. After parenting twin girls, now age 4, John and I were very nervous about adding child “#3” into the mix. But after having such a wonderful, complication-free pregnancy this time, we were sure the birthing process would run smoothly and giving birth to one child this time would be a pleasure.

My due date was March 27th, 2002; however Josh decided to come into the world a little early, the morning after my 35th birthday on March 19th, 2002. I was told I was GBS positive one week prior to my delivery.

My labor started around 4:00 a.m. the 19th of March and it came on pretty strong. My waters broke about 4:45 a.m. which is when I called Vassar Hospital to let them know about the GBS and that I would be on my way. We got to the hospital at about 5:30 a.m. and at this point the contractions were so bad that I could not speak or move. Much different this time than with the girls because I was induced and the labor process seemed more gradual and much longer. I pushed my son out within 15 minutes and Joshua was born at 7:14 a.m."

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HELLP Syndrome

HELLP syndrome (hemolysis, elevated liver enzymes, low platelets) is a serious complication of preclampsia that was first described by Pritchard et al, although Weinstein coined the term HELLP syndrome in 1892. Among women with severe preclampsia, 6% will manifest with one abnormality suggestive of HELLP syndrome (usually elevated liver enzymes or low platelets), 12% will develop two abnormalities, and about 10% will manifest with all three abnormalities. HELLP syndrome can manifest at any time during pregnancy and the puerperium but (like preclampsia) is rare before 20 weeks’ gestation. One third of all cases of HELLP syndrome occur postpartum, and only 80% of such patients were diagnosed with pre-eclampsia before delivery.

CLINICAL MANIFESTATIONS AND DIAGNOSIS

Although parturients with HELLP syndrome may be asymptomatic, 80% report right upper quadrant pain and worsening edema. Not all women with HELLP syndrome have hypertension or proteinuria. Indeed, 20% of patients with HELLP syndrome have a maximum blood pressure less than 140/90 mmHg, and 6% do not have significant proteinuria at the time of diagnosis. Other clinical conditions that should be considered in women with features suggestive of HELLP syndrome include hemolytic uremic syndrome, thrombotic thrombocytopenic purpura and acute fatty liver. The definitive laboratory criteria for the diagnosis of HELLP syndrome remain to be validated prospectively. However, the laboratory criteria most commonly used are those defined by Sibai. Sabai defined hemolysis as the presence of an abnormal peripheral smear with schisocytes, serum lactate dehydrogenase (LDH) more than 600 U/L, and total bilirubin more than 1.2 mg/dL; elevated liver enzymes as serum aspartate aminotransferase more than 70 U/L (>3 standard deviations about norm) and LDH more than 600 U/L; and low platelet count as less than 100,000/mm³. Based on the severity of the thrombocytopenia, Martin et al further categorized HELLP syndrome into three classes. Class 1 is defined as a platelet count less than 50,000/mm³, class 2 as a platelet count 50,000 to 100,000/mm³ and class 3 as a platelet count more than 100,000/mm³. Although this classification does appear to correlate to some degree with the prognosis and speed of resolution, it is not widely accepted.

ETIOLOGY AND PATHOPHYSIOLOGY

Like preclampsia, endothelial dysfunction, with resultant activation of the intravascular coagulation cascade, has been proposed as the central pathogenesis of HELLP syndrome. However, unlike preclampsia, HELLP syndrome occurs more often in whites, in multipara, and in women older than 35 years. Some investigators regard HELLP syndrome as an entirely distinct disease entity from preclampsia.

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When preclampsia is complicated by HELLP syndrome the maternal and perinatal death rates are significantly increased. Reported maternal death results most often from liver rupture, DIC, acute renal edema, carotid thrombosis, and cerebrovascular accident. Perinatal death is related most closely to complications of prematurity, fetal growth abruption. Reported perinatal death rates are 7.7-60%. Delayed diagnosis and delayed or inappropriate treatment are commonly cited as reasons from the poor overall prognosis associated with HELLP syndrome. Early identification of this syndrome, coupled with prompt and appropriate intervention, can significant reduces maternal and perinatal death and complication rates. Accordingly, parturients with HELLP syndrome should ideally be managed in a tertiary care facility.

MANAGEMENT

Stabilization of the mother’s conditions and assessment of fetal well being are the first responsibilities of management for parturients with HELLP syndrome. Seizure prophylaxis should be administered in the form of parenteral magnesium sulfate. If the pregnancy is less than 34 weeks, antenatal corticosteroids should be given to enhance fetal lung maturation. With few exceptions, immediate delivery is indicated, irrespective of gestational age. The decision of whether to delay delivery for 48 hours to complete a full course of antenatal corticosteroids should be individualized. Immediate delivery does not necessarily mean cesarean delivery. However, if the pregnancy is remote from term (<32 weeks) and the cervix is unfavorable, an elective cesarean delivery is a reasonable option. Because the incidence of hemotoma formation after cesarean delivery in women with HELLP syndrome may be as high as 20%, it may be prudent to place one or more subfascial and/or subcutaneous drains at the time of surgery, especially if there is evidence of significant intraoperative oozing or severe thrombocytopenia (<50,000/mm³). The drains can be removed electively in 24 to 48 hours. If the gestational age is more than 34 weeks, induction of labor can be initiated with or without cervical ripening, if indicated. It is our usual practice to check coagulation test results as well as hepatic and renal function test results every 6 hours until delivery, and then daily when stable. Several specific therapeutic maneuvers have been proposed in an effort to cure or alleviate HELLP syndrome. These include, among others, plasma volume expansion (using crystalloid or albumin), thrombolytic agents (low-dose aspirin, dipyridamole, heparin, antithrombin III, prostacyclin/thromboxane synthetase inhibitors), immunosuppressive agents (corticosteroids), exchange plasmapheresis, and dialysis. Magann et al reported that antepartum dexamethasone administration to women with HELLP syndrome significantly increased maternal platelet count, decreased serum alanine aminotransferase and LDH, increased maternal urine output, and resulted in a longer entry-to-delivery interval compared with women who did not receive corticosteroids.

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A subsequent study by the same group from the University of Mississippi Medical Center reported that dexamethasone was more effective than betamethasone in the antepartum “treatment” of HELLP syndrome. Of note, the dose of dexamethasone recommended in these studies for antepartum treatment of HELLP syndrome (12 mg q12h until delivery) is significantly higher than that recommended by the National Institutes of Health or the American College of Obstetricians and Gynecologists (ACOG) for promotion of fetal lung maturity (6 mg q12h for 48 hours). Moreover, corticosteroid administration in these studies was by the intravenous rather than the intramuscular route, as recommended by the National Institutes of Health and ACOG. The effect of large doses of intravenous corticosteroids on fetal adrenal function and fetal development is not known. As such, expectant management and antepartum “treatment” of HELLP syndrome with large doses of corticosteroids is not universally accepted.

In addition to antepartum corticosteroids, Magann et al have also reported on the use of postpartum intravenous corticosteroids (10 mg q12h for two doses followed by 5 mg q12h for two doses) to accelerate the reversal of HELLP syndrome. Although the fetal effect of high-dose corticosteroids is no longer a concern postpartum, larger randomized clinical trials are needed to verify the efficacy of postpartum corticosteroid therapy for HELLP syndrome. With or without corticosteroids, the vast majority of women with HELLP syndrome will recover within 96 hours of delivery.

FUTURE PREGNANCIES

The reported risk of recurrent HELLP syndrome in a subsequent pregnancy ranges from 3% to 27%. Future pregnancies are also at increased risk of other adverse events, including other manifestations of preclampsia, preterm delivery, fetal growth restriction, placental abruption, and cesarean delivery. The overall risk of such complications is 19-43%.

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Hang In There
Spring Is Coming!

Joshua
on ECMO
March 2002

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Internet Sites for Parents
www.cpr-ecc.org/search_ecc.asp: American Heart Association web site to locate a CPR course in your community.
http://www.pediatrics.wisc.edu/childrenshosp/parents_of_preemies: For Parents of Preemies, answers to commonly asked questions written by neonatologist Jane Brazy, M.D. There is also a Spanish
http://www.pediatrics.wisc.edu/childrenshosp/Preemie_Parent_Sp/spindex.html: Una place created for parents and loved ones of premature infants, and women experiencing a high risk or complicated pregnancy.
http://www.preemie-l.org: Parents of Premature Babies Inc. (Preemie-L)
A non-profit foundation supporting families with children born six weeks or more before due date.
http://kingproductions.com: Tommy's Cyber Nursery
Preemie Web - Front Door... Links to preemie Info, other preemie websites, discussion groups, medical information.
http://www.wcmc.com: Westchester Medical Center Homepage
http://www.nymc.edu/neonatology: New York Medical College, Division of Neonatology web page
http://www.modimes.org: March of Dimes organization whose goal is to reach the day when all babies are born healthy.
( The content of each website is the responsibility of the webmaster.)

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