Blood Disorders in Women
Challenges Across the Lifespan:
Hemoglobinopathies

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Memorial Regional Hospital, Hollywood, FL
and Chief Medical Officer
Sickle Cell Disease Association of America, Inc.
Hemoglobinopathy Across the Lifespan (Sickle Cell Disease)

<table>
<thead>
<tr>
<th></th>
<th>Infancy (0-6 months)</th>
<th>Early Childhood (6 months-3 years)</th>
<th>Pediatric (2-18 years)</th>
<th>Adolescent Young Adult (16-22 years)</th>
<th>Adult (≥ 18 years)</th>
<th>Older Adults</th>
</tr>
</thead>
<tbody>
<tr>
<td><strong>Mild anemia</strong></td>
<td>Pain</td>
<td>Pain</td>
<td>Anemia</td>
<td>Anemia</td>
<td>Anemia</td>
<td>Pain</td>
</tr>
<tr>
<td><strong>Penicillin</strong></td>
<td>Anemia</td>
<td>Anemia</td>
<td>Stroke</td>
<td>Stroke</td>
<td>Stroke</td>
<td>Stroke</td>
</tr>
<tr>
<td><strong>Aplastic crises</strong></td>
<td>Stroke</td>
<td>Priapism (Erection of Penis from SCD)</td>
<td>Priapism (Erection of Penis from SCD)</td>
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</tr>
<tr>
<td><strong>Hand foot syndrome</strong></td>
<td>Delayed puberty</td>
<td>Hearing loss (high frequency)</td>
<td>Lung* (ACS, PHT, Asthma)</td>
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<td>Lung* (ACS, PHT, Asthma)</td>
</tr>
<tr>
<td><strong>Spleenic sequestration</strong></td>
<td>Gallstones</td>
<td>Gallstones</td>
<td>Bone **(AVN)</td>
<td>Bone **(AVN)</td>
<td>Bone **(AVN)</td>
<td>Bone **(AVN)</td>
</tr>
<tr>
<td><strong>Jaundice</strong></td>
<td>*Pulmonary Hypertension (PHT)</td>
<td><strong>Avascular Necrosis</strong></td>
<td><strong>Avascular Necrosis</strong></td>
<td><strong>Avascular Necrosis</strong></td>
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</tr>
</tbody>
</table>

**Menstrual Issues, Pain Crisis, Contraception, Pregnancy**

**Transitions All Ages**
- Mental Health
- Academic Progress & Achievement
- Quality of Life
- Employment & Career
- Transcranial Doppler (head ultrasound study for risk of stroke)
- Hydroxyurea
- Transfusions/Iron Overload
- Associated conditions (Infections, Penicillin Prophylaxis)
- Newborn Screening/Sickle Cell Trait Status

**Infections**

**Neurocognitive Testing**

**Health Services/Medical Home/Access**
- Nutrition
- Mortality
- Transplant
- Insurance
UNINTENDED PREGNANCY

• Safe and effective contraceptives
  - Depo-Provera
  - Combined OCP

• Contraceptives and concomitant effect on painful crisis
  - Amelioration of painful crisis associated with menstruation
Are painful episodes in women with SCD associated with their menstrual cycles?


- 52 women, all SCD phenotypes
- 34% reported acute painful episodes with their periods

Yoong WC and Tuck SM. Menstrual pattern in women with sickle cell anemia and its association with sickling crises. 2002.

- 42 women, SS only
- 37% with painful episodes with periods
Effect of Depo-Provera or Microgynon on the Painful Crises of Sickle Cell Anemia Patients

de Abood M et al, Contraception 1998; 56:313-6

• Prospective study
• To compare the intensity and frequency of acute painful episodes in three groups of women with SCD
• Three study groups
  – combined OCP (levonorgestrel (0.15 mg) + ethinyl estradiol (0.03 mg))
  – Depot medroxyprogesterone acetate (DMPA) injectable (150mg monthly x 3 months then every 3 months
  – surgically sterilized
• Eligibility – SS, one painful episode/month
• Study period: 12 months
• Group 1 – 13, Group 2 - 14, Group 3 - 16
Variation in Pain Frequency in Three Study Over 12-month Study Period
PREGNANCY

• Preconception counseling and access to family planning programs are of particular importance to women with sickle cell disease (SCD) given studies that suggest that pregnancies in women with SCD are high-risk
## Complications of SCD During Pregnancy

<table>
<thead>
<tr>
<th>Thromboembolic Events</th>
<th>OR</th>
<th>95% CI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stroke</td>
<td>2.0</td>
<td>0.6 - 6.9</td>
</tr>
<tr>
<td>Cerebral vein thrombosis</td>
<td>4.9</td>
<td>2.2 – 10.9</td>
</tr>
<tr>
<td>Pulmonary embolism</td>
<td>1.7</td>
<td>0.9 – 3.1</td>
</tr>
<tr>
<td>Deep vein thrombosis</td>
<td>2.5</td>
<td>1.5 – 4.1</td>
</tr>
</tbody>
</table>

## Complications of SCD During Pregnancy

<table>
<thead>
<tr>
<th>Infections</th>
<th>OR</th>
<th>95% CI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pneumonia</td>
<td>9.8</td>
<td>8.0 – 12.0</td>
</tr>
<tr>
<td>Pyelonephritis</td>
<td>1.3</td>
<td>1.0 – 1.8</td>
</tr>
<tr>
<td>Asymptomatic bacteruria</td>
<td>6.8</td>
<td>3.1 – 14.9</td>
</tr>
<tr>
<td>Genitourinary infection</td>
<td>2.3</td>
<td>1.9 – 2.7</td>
</tr>
<tr>
<td>Sepsis</td>
<td>6.8</td>
<td>4.4 – 10.5</td>
</tr>
<tr>
<td>SIRS</td>
<td>12.6</td>
<td>2.1 – 13.6</td>
</tr>
</tbody>
</table>

## Complications of SCD During Pregnancy

<table>
<thead>
<tr>
<th>Obstetrical Complications</th>
<th>OR</th>
<th>95% CI</th>
</tr>
</thead>
<tbody>
<tr>
<td>Preeclampsia</td>
<td>1.2</td>
<td>1.2 – 1.3</td>
</tr>
<tr>
<td>Eclampsia</td>
<td>3.2</td>
<td>1.8 – 6.0</td>
</tr>
<tr>
<td>Abruption</td>
<td>1.6</td>
<td>1.2 – 2.1</td>
</tr>
<tr>
<td>Antepartum bleeding</td>
<td>1.7</td>
<td>1.2 – 2.2</td>
</tr>
<tr>
<td>Preterm labor</td>
<td>1.4</td>
<td>1.3 – 1.6</td>
</tr>
<tr>
<td>Fetal growth restriction</td>
<td>2.2</td>
<td>1.8 – 2.6</td>
</tr>
<tr>
<td>Intrauterine fetal demise</td>
<td>1.1</td>
<td>0.8 – 1.7</td>
</tr>
<tr>
<td>Postpartum hemorrhage</td>
<td>0.5</td>
<td>0.3 – 0.6</td>
</tr>
</tbody>
</table>

Duke Experience

158 pregnancies

- 111 live (26% LBW)
- 3 stillbirths
- 43 abortions
- 1 ectopic

- 28 spontaneous
- 15 induced

Duke Experience

- Mean gestational age 36 weeks
- 5/158 with DVT
- 1/158 with PE
- Overall VTE = 4%
- Increasing pain medication associated with lower birth weight and longer hospital stay

Health Policy

Fair and Equitable for Women and Girls?
Women have unique health care needs, and often make health care decisions for their families. The law offers important benefits for women and their families.
CONTRACEPTION AND FAMILY PLANNING

The Patient Protection and Affordable Care Act – the health insurance reform legislation passed by Congress and signed into law by President Obama on March 23, 2010 – will help make prevention affordable and accessible by requiring health plans to cover recommended preventive services without charging a deductible, copayment or co-insurance.
The U.S. Department of Health and Human Services charged the Institute of Medicine (IOM) with reviewing what preventive services are important to women’s health and well-being and then recommending which of these should be considered in the development of comprehensive guidelines.
Actions Taken

Women’s Preventive Services Recommended by IOM to be Covered Under Affordable Care Act
The IOM defined preventive health services as measures—including medications, procedures, devices, tests, education and counseling—shown to improve well-being, and/or decrease the likelihood or delay the onset of a targeted disease or condition.
**TABLE S-1 Continued**

<table>
<thead>
<tr>
<th>Preventive Service</th>
<th>USPSTF Grade</th>
<th>Supporting Evidence</th>
<th>Recommendations</th>
</tr>
</thead>
<tbody>
<tr>
<td>Contraceptive methods and counseling</td>
<td>Not Addressed</td>
<td>The evidence provided to support a recommendation related to unintended pregnancy is based on systematic evidence reviews and other peer-reviewed studies, which indicate that contraception and contraceptive counseling are effective at reducing unintended pregnancies. Current federal reimbursement policies provide coverage for contraception and contraceptive counseling, and most private insurers also cover contraception in their health plans. Numerous health professional associations recommend family planning services as part of preventive care for women. Furthermore, a reduction in unintended pregnancies has been identified as a specific goal in <em>Healthy People 2010</em> and <em>Healthy People 2020</em>.</td>
<td>Recommendation 5.5 The committee recommends for consideration as a preventive service for women: the full range of Food and Drug Administration-approved contraceptive methods, sterilization procedures, and patient education and counseling for women with reproductive capacity.</td>
</tr>
</tbody>
</table>
IOM Recommendations

• Improved screening for cervical cancer, counseling for sexually transmitted infections, and counseling and screening for HIV;

• A fuller range of contraceptive education, counseling, methods, and services so that women can better avoid unwanted pregnancies and space their pregnancies to promote optimal birth outcomes;

• Services for pregnant women including screening for gestational diabetes and lactation counseling and equipment to help women who choose to breastfeed do so successfully;

• At least one well-woman preventive care visit annually for women to receive comprehensive services; and

• Screening and counseling for all women and adolescent girls for interpersonal and domestic violence in a culturally sensitive and supportive manner.
Affordable Care Act – 22 Preventive Services for Women, Including Pregnant Women (healthcare.gov)

Eight Preventive Services must be covered with no cost-sharing in plan years starting on or after August 1, 2012 (Breastfeeding, Contraception, Domestic Violence, Gestational Diabetes, HIV, HPV, STI, Well woman visits)

**Contraception:** Food and Drug Administration-approved contraceptive methods, sterilization procedures, and patient education and counseling, not including abortifacient drugs
Women Can Receive **Preventive Care Without Copays**. Thanks to the Affordable Care Act, all Americans joining a new health care plan can receive recommended preventive services, like mammograms, new baby care and well-child visits, with no out-of-pocket costs. See a list of preventive services for women. (Preventive services benefits apply if you’re in a new health plan that you joined after March 23, 2010.) Learn about new **women's preventive care guidelines** issued August 1, 2011.
Women Pay Lower Health Care Costs. Before the law, women could be charged more for individual insurance policies simply because of their gender. A 22-year-old woman could be charged 150% the premium that a 22-year-old man paid. In 2014, insurers will not be able to charge women higher premiums than they charge men. The law takes strong action to control health care costs, including helping states crack down on excessive premium increases and making sure most of your premium dollars go for your health care.
Preconception Recommendations for Women with Sickle Cell Disease

• Encourage each woman, man, and couple to have a reproductive life plan. Advise or refer for advice, women with sickle cell disease who anticipate pregnancy and their partners that a pregnancy is high-risk

• As a part of primary care visits, provide risk assessment and educational and health promotion counseling to all women of childbearing age to reduce reproductive risk and improve pregnancy outcomes

• If the partner of a man or woman with sickle cell disease has an unknown SCD or thalassemia status, refer them for hemoglobinopathy testing

• After testing, refer couples who are at risk for having a potentially affected fetus/neonate for genetic counseling, i.e. neonatal withdrawal syndrome in newborns exposed to chronic narcotic use in-utero

• Inform or refer for education, women with SCD and their partners, about the additional fetal surveillance required during a pregnancy

• Inform or refer for information, women and their partners, about the increased risks to a woman’s health during pregnancy. These risks include an increased frequency of pain crises and an increased risk of thromboses, infections, preeclampsia and death relative to other women
• Provide counseling on full range of contraception options for women with Sickle Cell Disease

• Provide counseling on pregnancy health risk and contraindications to pregnancy

• Elimination of co-pays for preventive care removes an important barrier to women seeking and receiving care

• Sickle Cell Disease is not a contraindication to pregnancy

• Following appropriate, evidence-based preconception care guidelines is paramount
Thank you    Gracias    Merci