RED BLOOD CELL EXCHANGE IN SICKLE CELL DISEASE – A CONSENSUS CONFERENCE

at the ASFA 2015 Annual Meeting

ASFA
American Society for Apheresis

2015

SAN ANTONIO, TX

Grand Hyatt • May 5th, 2015

AN EDUCATIONAL AND NETWORKING FORUM
FOR PROFESSIONALS IN THE FIELD OF APHERESIS MEDICINE

Accreditation

Please note that the target audience, statement of need, program overview, educational objectives and faculty disclosures are provided in the ASFA 2015 Annual Meeting Final Program. Please visit the ASFA Registration Desk for more information.

PHYSICIANS

This activity has been planned and implemented in accordance with the Essential Areas and Policies of the Accreditation Council for Continuing Medical Education (ACCME) through the joint providership of the American Association of Blood Banks (AABB) and the American Society for Apheresis (ASFA). AABB is accredited by the ACCME to provide continuing medical education for physicians (Provider number 0000381). AABB designates this educational activity for a maximum of 7.75 of Category 1 credit toward the *AMA Physicians Recognition Award*™. Each physician should claim those credits that he/she actually spent in the activity.

FACULTY DISCLOSURE

Current ACCME guidelines state that participants in CME activities should be made aware of any affiliation or financial interest that may affect a speaker's presentation and/or discussion of off-label therapies. Each speaker was asked to complete a Faculty Disclosure Form. Written faculty disclosures will be provided to participants in the syllabus. Please report any undisclosed information on your evaluation form.

LIVE LEARNING CENTER

After the annual meeting, you will receive an email from AABB regarding the CME/CE certificate for the ASFA 2015 Annual Meeting. The e-mail will include instructions on how to print your Physician CME/CE certificates. To access the Live Learning Center, visit www.aabb.org > Professional Development > Live Learning Center.

CEU

ASFA is approved by the California Board of Registered Nursing, provider number 14122, as a provider of continuing nursing education programs. ASFA designates this event for a maximum of 7.75 contact hours.

CMLE

This continuing medical laboratory education activity is recognized by the American Society for Clinical Pathology (ASCP) as meeting the criteria for 7.75 of CMLE credit. ASCP CMLE credit hours are acceptable to meet the continuing education requirement for the ASCP Board of Registry Certification Maintenance Program.

DISCLOSURE OF CONFLICTS OF INTEREST

Faculty Name	Name of Commercial Interest	Relationship
Mark Gladwin	Bayer	Consultant, Grant/Research Support
	NIH/NHLBI	Grant/Research Support
	Aires	Grant/Research Support
	Institution for Transfusion Medicine	Grant/Research Support
	Hemophilia Center of Western PA	Grant/Research Support
	VitaBeet (Sl.)	Stock Shareholder (wife)
Keith Quirolo	Terumo BCT	Honoraria, Consultant
	AngioDynamics	Discussion of off-label/investigative uses

Consensus Conference Organizing Committee

Ravi Sarode, *MD, UT-Southwestern Medical Center, Chair of the Consensus Conference Organizing Committee*
Bruce Sachais, *MD, PhD, New York Blood Center*
Samir Ballas, *MD, Thomas Jefferson University*
Eileen Galvin Karr, *RN, BSN, HP(ASCP), Baystate Medical Center*
Alicia Garcia, *RN, HP(ASCP), Children's Hospital and Research Center Oakland*
Haewon Kim, *MD, Children's Hospital of Philadelphia*
Karen King, *MD, Johns Hopkins Hospital*
Lance Williams, *MD, University of Alabama at Birmingham*

Consensus Conference Program

TUESDAY, MAY 5, 2015 — LONE STAR BC

7:00 AM – 6:00 PM	*Speaker Services*	*Goliad*
7:00 AM – 7:45 AM	*Registration and Continental Breakfast*	*Escalator Landing Second Floor*
7:45 AM – 8:00 AM	**WELCOME AND INTRODUCTION** **Ravi Sarode,** *MD, UT-Southwestern, Chair of the Consensus Conference Organizing Committee* **Moderators: George Buchanan,** *MD, UT-Southwestern Medical Center and* **Mark Brecher**, *MD, Laboratory Corporation of America and University of North Carolina*	
8:00 AM – 9:00 AM	**TOPIC 1: PATHOPHYSIOLOGY OF SICKLE CELL DISEASE** **Martin H. Steinberg,** *MD, Boston University*	

	8:00 AM – 8:45 AM	**DISCUSSION POINTS**
		• Discuss the pathophysiology of the clinical sequelae of vaso-occlusion and vasculopathy.
		• Discuss how red blood cell exchanges and/or simple transfusions exert their clinical effect, with a focus on the following:
		  • Rheology/viscosity as the basis to optimize a patient's Hemoglobin S (HbS) and Hemoglobin levels
		  • Ischemia
		  • Reperfusion injury and end-organ damage
		  • Inflammation and hypercoagulable state
		  • Pain (acute and chronic)
		  • Priapism and ASPEN syndrome

	8:45 AM – 9:00 AM	Questions & Discussion

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9:00 AM – 10:00 AM	**TOPIC 2: ACUTE CHEST SYNDROME (ACS)** **Keith Quirolo,** *MD, UCSF Benioff Children's Hospital*	
	9:00 AM – 9:45 AM	**DISCUSSION POINTS** **Etiology:** • Discuss the possible etiologies of ACS, as well as cause-effect relationships and risk factors for both initial and recurrent episodes, with special consideration of the following: 1. Pulmonary hypertension 2. Interstitial lung disease **Diagnosis:** • Discuss the differential diagnosis of ACS, including relevant tests and clinical findings for preliminary (early), and definitive diagnosis to reduce morbidity and mortality, with a focus on ACS versus the following conditions: 1. Pulmonary embolus 2. Pulmonary infarction 3. Fat embolus **Treatment:** • Discuss the laboratory and/or clinical thresholds for treatment of ACS using the following strategies: 1. Simple transfusion • Is there any role/rationale for simple transfusion in acute ACS? • Is there any evidence or rationale for chronic transfusion therapy to prevent recurrent ACS? 2. RBCx • What are the acute treatment goals? (e.g. H&H, HbS, O2 Sats, etc.) • Are there any variables that affect the efficacy of the RBCx? (e.g. patient age, previous episodes of ACS, co-morbidities, lab values, etc.)
	9:45 AM – 10:00 AM	Questions & Discussion
10:00 AM – 10:25 AM	*Break*	
10:25 AM – 10:30 AM	**MODERATOR SUMMARY**	

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10:30 AM – 11:30 AM	**TOPIC 3: CEREBROVASCULAR ACCIDENTS (CVA)** **Michael DeBaun,** *MD, Vanderbilt University*	
	10:30 AM – 11:15 AM	**DISCUSSION POINTS** **Pathophysiology:** • Why are the cerebral arteries more prone to vasculopathy? **Treatment; Acute and Chronic Management/Prevention of CVA:** • During an acute stroke, what is the optimal management between simple transfusion and RBCx? **After an acute stroke, discuss chronic management to include the following:** • Is there any evidence to suggest superiority of RBCx versus simple transfusion? • What laboratory values (e.g. HbS) or imaging studies (transcranial doppler (TCD)) should be used to determine the necessity and frequency of prophylactic transfusion therapy after a stroke? • What are the treatment thresholds in children and adults? • Are higher thresholds of <50% for HbS safe? • Once TCDs are normal, is it safe to raise the hemoglobin to near normal levels? • What is the recommended duration of any chronic transfusion therapy for primary stroke prevention in children and adults? (i.e. 3 years versus lifelong) • What are the risk factors for recurrent CVA and Moyamoya syndrome?
	11:15 AM – 11:30 AM	Questions & Discussion
11:30 AM – 12:15 PM	**TOPIC 4: PULMONARY HYPERTENSION (PH)** **Mark Gladwin,** *MD, University of Pittsburgh*	
	11:30 AM – 12:00 PM	**DISCUSSION POINTS** **Diagnosis:** • Provide a brief overview of the prevalence of pulmonary hypertension (PH) and its morbidity/mortality in patients with sickle cell disease. • What are the optimal screening (e.g. Pulmonary Function Tests (PFTs)) and diagnostic tests (e.g. Tricuspid Regurgitant Jet (TRJ) velocity, Right Heart Catheterization (RHC), etc.) for PH in this patient population? • Is it possible to predict, by testing or clinical signs/symptoms, which patients will progress to PH? **Treatment:** • Is there a role for chronic transfusion therapy (either simple transfusion or RBCx) in the treatment or in the prevention of PH in at-risk patients? If so, discuss the rationale for specific treatment goals.

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11:30 AM – 12:15 PM	**TOPIC 4: PULMONARY HYPERTENSION *continued*** Mark Gladwin, *MD, University of Pittsburgh*	
	12:00 PM – 12:15 PM	Questions & Discussion
12:15 PM – 12:55 PM	*Lunch*	
12:55 PM – 1:00 PM	**MODERATOR SUMMARY**	
1:00 PM – 2:00 PM	**TOPIC 5A: RED CELL EXCHANGE PROCEDURE – STANDARD AND ISOVOLEMIC HEMODILUTION (IHD) RBCX FOR CHRONIC TRANSFUSION THERAPY** **TOPIC 5B: SELECTION OF RED CELLS** Haewon Kim, *MD, Children's Hospital of Philadelphia* Araba Afenyi-Annan, *MD, University of North Carolina – Chapel Hill*	
	1:00 PM – 1:35 PM	**TOPIC 5A: RED CELL EXCHANGE PROCEDURE – STANDARD AND ISOVOLEMIC HEMODILUTION (IHD) RBCX FOR CHRONIC TRANSFUSION THERAPY**
	Haewon Kim, *MD, Children's Hospital of Philadelphia*	
	Discussion Points • What are the advantages and disadvantages of each procedure? • What are the long-term benefits of each procedure? • Should IHD-RBCx procedures become the "standard of care" for patients requiring long-term transfusion therapy? • What are the contraindications for IHD-RBCx procedures? • What are the optimal target levels for Hemoglobin/Hematocrit and HbS after RBCx? • Should these targets be adjusted based on the patient's pre-procedure hematocrit and serum ferritin level? • What are the challenges in performing RBCx in small children?	
	1:35 PM – 1:45 PM	**TOPIC 5B: SELECTION OF RED CELLS**
	Araba Afenyi-Annan, *MD, University of North Carolina - Chapel Hill*	
	Discussion Points • What evidence/guidelines exist for using phenotype matched units during RBCx to prevent alloimmunization? • What is the cost-benefit ratio of phenotype matching? • Are there any advantages of using fresh blood (<7 days old), either for acute or chronic RBCx?	
	1:45 PM – 2:00 PM	Questions & Discussion

TUESDAY, MAY 5, 2015 — LONE STAR BC

2:00 PM – 2:45 PM	**TOPIC 6: TECHNICAL AND NURSING ASPECTS** **Cathy Hulitt,** *RN, Children's Hospital of Philadelphia*	
	2:00 PM – 2:30 PM	**Discussion Points** • What are the access options for acute and chronic RBCx? • What are the options for various implantable ports (Vortex and Sport) with a focus on advantages/disadvantages, accessing issues, durability, effects on patient lifestyle, etc.? • What are the optimal anticoagulants and techniques for flushing and locking access lines? • The choice of sedation versus no sedation in children.
	2:30 PM – 2:45 PM	Questions & Discussion
2:45 PM – 2:55 PM	*Break*	
2:55 PM – 3:00 PM	**MODERATOR SUMMARY**	
3:00 PM – 5:00 PM	**CONSENSUS DELIBERATION**	
7:00 PM – 10:00 PM	**ASFA BOARD OF DIRECTORS AND RED BLOOD CELL EXCHANGE IN SICKLE CELL DISEASE CONSENSUS CONFERENCE SPEAKERS DINNER** *(BY INVITATION ONLY, OFFSITE)*	

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