Vincent’s Circle of Life Sense

Like the Academy Awards it is all about performances. We have directors, producers, writers, composers, musicians, actors and their roles. We have imaginations, innovations, pioneers and time for the agonies and heartaches about prior cases beyond our control only to motivate and drive creative thoughts so all enchanted dreams will come true. Put them together and what do you got? (Bibbidi bobbidie boo), rather, a team and togetherness. It’s all about teamwork, and we are all in this together. Allow me to refer you to a couple previously published Links articles: one on Collaboration, Conformity and Consensus (October 2011, p 15-16) and the other on Linking it all Together (April 2012). Of course Benjamin Franklin comes to mind as he reminds us "we must indeed, all hang together or most assuredly, we shall all hang separately." Over the last three decades we have witnessed the impressive and marvelous progress in the world of pulmonary hypertension and pediatric heartaches with new classes of medications and novel ways to use mechanical devices.

The Wonderful World of Disney is obviously part of shaping our minds at least through our children. And over the last three decades adults have learned a great deal from children about safety, using seatbelts and helmets, saying no to cigarettes, to drugs, to booze, to sugar and to bullying and saying yes to exercise, to vaccines and to other healthful habits. As such, Murali Chakinala shows us the way with the evolution of Pulmonary Hypertension Care Centers, a major lesson learned from the more than a half century experience from the Cystic Fibrosis Foundation, a lesson for adults from children or the Wonderful World of Pediatrics. The performances (fewer symptoms, longer distances walked and better quality) in the world of pulmonary hypertension as shared from their articles by Veronica Franco, Manreet Kanwar and Julia Estes show that the lives, lifestyles and symptomatic improvements in these afflicted patients today were all but a dream 30 years ago.

The Next Frontier (Sharon Chen and Beth Kaufman), the Challenges (Marc Schecter) and the Lessons Learned (Janet Scheel) come from taking what we know is best for adults and courageously applying to our little ones who we know are not just merely little adults but at least gives us the warmth, confidence and sense of well-being to push the boundaries from in utero to octogenarians. It will be only a matter of time that the adult world of transplantation, assist and supportive devices, and perhaps other forms of replacements and innovative strategies will learn from the innocence of our children and our fantasies to once again complete the cycle = adults - children - adults - children ... Hakuna matata.

Vincent Valentine, MD
Links Editor-in-Chief
In The Spotlight: ISHLT 2014 in Sunsational San Diego!

Featuring Meeting Highlights in Pulmonary Hypertension and Pediatric Transplantation

At ISHLT 2014, there will be a number of symposia with primary content of interest in pulmonary hypertension as well as pediatric transplantation. Find out which sessions will be of most interest to you, and plan your trip to San Diego TODAY!

Focus on Pulmonary Hypertension:

We are very pleased to present an outstanding program at ISHLT 2014 that covers a diverse range of innovations and controversial topics in Pulmonary Hypertension. There are four Pre-Meeting Symposia, three Sunrise Sessions, and one Concurrent session that will highlight the latest clinical consensus statements, discuss the emerging sciences, and pose questions to the experts and audiences for discussion and feedback. These sessions will be of keen interest to all members of ISHLT engaged in the care of patients with cardiopulmonary disease and right heart failure.

PRE-MEETING SYMPOSIUM 2: Pulmonary Hypertension: You Can't Ignore It any Longer (Thursday, April 12, 8:00 AM - 10:00 AM)
To kick off the meeting, this session will tackle the controversial area of managing patients with lung disease and pulmonary hypertension, WHO Group III, focusing on patients with ILD, COPD and sarcoidosis.

PRE-MEETING SYMPOSIUM 8: CTEPH: Busting the Clot (Thursday, April 12, 10:15 AM - 12:15 PM)
What better place to focus on all the latest exciting developments in chronic thromboembolic pulmonary hypertension (CTEPH) than San Diego! This session features both medical and surgical aspects in managing CTEPH patients with presentations highlighting the emerging new treatment, riociguat, as well as a debate to consider exercise-induced PH as an indication for pulmonary thromboendarterectomy.

PRE-MEETING SYMPOSIUM 14: Treating All Stages of Severe PH: Are We Doing it Right? (Thursday, April 12, 2:00 PM - 4:00 PM)
This session will discuss the emerging basic sciences focusing on mechanisms and management approaches for patients with severe PH and right heart failure, from pulmonary vasodilators to mechanical supports. We are also very excited to present to you the ISHLT and World Symposium on PH (WSPH) Consensus Initiative on PH and Left Heart Disease in Pre-Meeting Symposium 20, which will present discussions asking what are the important hemodynamic variables in those with advanced heart failure and PH?

CONCURRENT SYMPOSIUM 25: A Lifecycle Journey in Pulmonary Hypertension (Friday, April 11, 10:30 AM - 12:00 PM)
This session will present the lifecycle journey of a patient facing PAH, focusing on clinical milestones and challenges that face these patients from diagnosis to advanced stages of disease.

**SUNRISE SYMPOSIUM 2: Under Too Much Pressure (Friday, April 11, 7:00 AM - 8:00 AM)**
This is meant to be a multidisciplinary symposium (cardiology, pulmonary, PH) discussing management of challenging patients who have complicating pulmonary hypertension. Three cases will be presented by junior faculty members. The case presentations will include challenges to the discussants (in an open forum) regarding next steps in evaluation, management, etc of pulmonary hypertension.

**SUNRISE SYMPOSIUM 7: The Effects of Prostaglandin Therapy in PAH: The Seen and Unseen Risk/Benefit Profile (Saturday, April 12, 7:00 AM - 8:00 AM)**
This session explores all the effects, both seen and unseen, in patients associated with parenteral prostanoid treatments. We are also very pleased to bring you a "How To" session, featuring oft asked questions regarding PA catheters in our Sunrise Symposium 12. You will get to engage in discussions regarding how to do volume loading, when to use vasodilator testing, and how to utilize exercise RHC and what the results mean.

**PULMONARY HYPERTENSION SCIENTIFIC COUNCIL MEETING (Saturday, April 12, 12:05-12:55 PM)**

**CLOSING PLENARY (Sunday, April 13, 9:30 AM - 11:45 AM)**
The grand finale, during the Closing Plenary session, will feature a debate you don't want to miss: Stop Treating Secondary PH Right Now! See which side you will vote for! We look forward to seeing you all in San Diego!

**Focus on Pediatric Transplantation:**

For members interested in pediatric topics, ISHLT 2014 promises to be busy and enlightening. In a series of Thursday Pre-Meeting Symposia, you will hear experts discuss preparing pediatric patients for transition to adult care, the evolution, the biology and the limits of ABO incompatible heart transplantation, Developing a pediatric VAD program, and State of the art update on infectious diseases in pediatric thoracic transplantation. And if you have ever wondered 'Why not infant lung transplantation?', a Sunrise Symposium during the meeting is planned just for you. And finally, a new Concurrent Symposium has been added recently to the program to address patient selection issues for pediatric thoracic transplantation.

**PRE-MEETING SYMPOSIUM 6: Joint ISHLT/IPTA Symposium: Here They Come: Preparing Pediatric Patients For Transition To Adult Care (Thursday, April 12, 8:00 AM - 10:00 AM)**
Transition is defined as “the process by which adolescents and young adults with chronic childhood illnesses are prepared to take charge of their lives and their health in adulthood”. Effective transition programs have the potential to decrease morbidity and mortality associated with transfer of care and can improve quality of life. This session will discuss issues essential to successful transition of pediatric patients to adult care, including patient and family challenges, as well as potential strategies/interventions to meet these challenges.
PRE-MEETING SYMPOSIUM 11: Crossing Clinical Barriers on the Wings of Science: Evolution of ABO Incompatible Heart Transplantation (Thursday, April 12, 10:15 AM - 12:15 PM)
This session has multiple purposes. It is intended to show how scientific concepts are developed for clinical application. It will show the history of ABO incompatible heart transplantation, but from the point of view of the designer of therapy. In so doing, it will delineate some of the technical details in the path of a very significant advance in transplantation from idea to clinical reality. In so doing, we hope that this session will be both informative, and inspirational. We intend for this session to stimulate the thought processes necessary to take other burgeoning ideas from bench to bedside by giving a better understanding of the processes involved in translating science to our patients.

PRE-MEETING SYMPOSIUM 18: Developing A Pediatric VAD Program (Thursday, April 12, 2:00 PM - 4:00 PM)
This session offers the following objectives:
1. To understand and review what is required to develop a VAD program at a pediatric center.
2. To review the outcomes of children discharged home on implantable VAD support
3. To understand important pediatric-specific medical and social obstacles impacting home VAD
4. To discuss the future of VAD support for children.

PRE-MEETING SYMPOSIUM 23: State of the Art Update on Infectious Disease Issues in Pediatric Thoracic Transplantation (Thursday, April 12, 4:15 PM - 6:15 PM)
The infectious disease sessions at ISHLT traditionally have focused on adults rather than children. Some of the pediatric responses to infections are quite disparate from those of adults. This session will provide a state of the art update based on the latest data in pediatrics.

PEDIATRIC TRANSPLANTATION SCIENTIFIC COUNCIL MEETING (Saturday, April 12, 12:05-12:55 PM)

CONCURRENT SYMPOSIUM 30: Controversies in Listing Children for Thoracic Organ Transplant (Saturday, 4/12/2014 2:00 PM - 3:30 PM)
This recently added session will address patient selection issues for pediatric thoracic transplantation. A great group of speakers has been assembled to stimulate the conversation. We hope this conversation will be the catalyst for a consensus conference that will provide cohesive guidelines for the thoracic transplant community.

SUNRISE SYMPOSIUM 14: Why Not Infant Lung Transplantation? (Sunday, April 13, 7:00 AM - 8:00 AM)
Less than 100 infant lung transplants have been performed in the world in the last 25 years whereas over 100 infant heart and liver transplants are performed each year. This disconnect suggests that a shortage of organs does not explain this result. Chronic respiratory or cardiopulmonary failure is not rare. A recent report indicates long-term outcome is at least as good for infant lung transplant recipients as for older recipients. This session will explore the conundrum.
Our understanding and management of pulmonary arterial hypertension (PAH) has advanced tremendously over the last 30 years. Numerous vasomodulating therapies have been developed, and their widespread use has been associated with longer survival and improved quality of life. Even though PAH remains a rare disease with challenging therapies, the delivery of health-care in PAH has been transformed from experts at tertiary care centers to a broad spectrum of providers with varying degrees of expertise, leading to non-uniformity of care. Concomitantly, PAH-specific therapy has been applied to an increasingly diverse population of patients with PH. As a result, early access to expert centers and assurances of optimal patient-care have become relevant concerns.

Recent publications have shed light on these challenges. In the RePHerral Study from 3 large university-based tertiary care referral centers in the US, 98 of 140 referred patients had been assigned a diagnosis of PAH before referral, but 32 (33%) were subsequently determined to be misdiagnosed. Forty-two patients were started on PAH-specific medications prior to referral, and 24 of these therapies were contrary to published guidelines. Fifty-nine patients had not had a pre-referral right heart catheterization [1]. Additional literature suggests that patients followed outside of a referral center (compared with the individuals already under the referral center’s care) are treated with oral therapies longer, are more compromised and more likely to need urgent initiation of parenteral prostanoids, and have lower survival rates even after prostanoids are initiated [2]. Evidence from the REVEAL registry demonstrates that a substantial number of patients in functional class III or IV within 6 months of death had not received parenteral prostanoid at the time of death, suggesting under-utilization of the most potent and effective class of therapies [3]. These reports intimate the perception of inaccurate diagnosis of PAH, untimely referral to expert centers, and inappropriate utilization of advanced therapies.

Two years ago, the PHA and its Scientific Leadership Council (SLC) spawned an initiative to address these challenges facing the PH community and created the Pulmonary Hypertension Comprehensive Care (PHCC) Committee. The initiative’s mission statement is to establish a program of accredited centers with expertise in pulmonary hypertension that aspires to improve overall quality of care and ultimately improve outcomes of patients with pulmonary hypertension, particularly pulmonary arterial hypertension, a rare and life-threatening disease.

The PHCC Committee studied and patterned the program after the successful Cystic Fibrosis Foundation Accredited Care Centers. The over-arching objectives of the PHCC program are to improve overall care and long-term patient outcomes, which can be accomplished through several interlocking components:
• Increasing disease awareness  
• Improving access to expert care  
• Raising the level of care at ALL centers through increased adherence to published guidelines and consensus statements  
• Providing a ‘blue print’ to prospective programs for becoming PH Care Centers  
• Fostering collaboration among expert Centers for managing individual patients and cultivating new research opportunities in the field  
• Conducting center-specific and national quality improvement projects with the aid of a national patient registry

The design for two types of Centers (i.e. Centers of Comprehensive Care and Regional Clinical Programs) is a central feature of the program that should maximize the eventual number of PHCCs across the country and enhance access to expert care. Both designations will be promoted as PHA-accredited PHCCs and will have to meet their respective criteria through the same application process and evaluation method. Both will have to broadly satisfy several categories of criteria, including Center Director, Center Coordinator, Program Staff/Support Services, Facilities, and Research (CCC only) [visit www.phassociation.org/phcarecenters]. Although inclusivity is emphasized, the criteria and the accreditation program still must adhere to standards for selecting PHCCs, so that the designation represents a tangible achievement and conveys meaningful information to relevant stakeholders.

The PHCC initiative has generated tremendous interest. At the cusp of launching this much needed grass-roots program, it is vital to appreciate the enormity of the project and its potential consequences without becoming paralyzed by fear and uncertainty. For the sake of our patients, the PH community needs to find the courage and perseverance to forge ahead!

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• Consulting:  
  Actelion, Gilead, United Therapeutics  
• Speaker’s Bureau:  
  Gilead, United Therapeutics

References:


Oral Prostacyclins: Our Newest Medication for Pulmonary Arterial Hypertension

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Pulmonary arterial hypertension (PAH) is a progressive, fatal disease that leads to right sided heart failure. Thirty years ago, adults diagnosed with PAH could expect to live less than 3 years, and the therapies limited to nonselective vasodilators (calcium channel blockers) and warfarin. Today, the physician has a number of choices and continues to expand with three new medications approved in 2013.

Prostacyclins, the first approved therapy, are still utilized and remain one of the main classes of PAH therapy. Currently, continuous SQ / IV formulations are typically utilized in severe PAH and patients with functional class III or IV. There is growing evidence that earlier use may benefit patients with mild to moderate disease [1]. Nevertheless, IV prostacyclins are not consistently used in the most seriously ill patients. Unfortunately they are often started late or not at all with ~ 50% of patients with PAH were not using prostacyclins at the time of their death [2]. In December 2013, the FDA approved an oral treprostinil, (Orenitram) for the treatment of PAH in WHO Group I patients. This approval marks the first time that the FDA has approved an orally administered prostacyclin analogue for PAH.

The FREEDOM-M study [3], evaluated oral treprostinil in newly diagnosed patients not yet treated with PAH specific medications. The study demonstrated that patients receiving treprostinil twice daily improved their median six-minute walk distance (6MWD) by +23 meters [p=0.013] as compared to patients receiving only placebo. The maximum dose was determined by tolerability. Side effects were similar to those of other prostacyclins – headache, nausea, diarrhea, hypotension, inhibition of platelet aggregation and risk of bleeding.

The approval of this medication provides potential significant advantages in the care for patients with PAH:

- Ability to provide the benefits of prostacyclin therapy to a broader population, as a consequence of the easier-to-use oral administration, without the risk of infection or pain at infusion site as seen with continuous IV and SQ formulations.
- Superior acceptance of its use by patients as well as by the medical community, resulting in earlier utilization of prostacyclin therapy in the course of the disease.
- Provides an additional medication available for the treatment of this complex and still fatal disease.
• Gradual titration of medication allowed based on tolerability. Several doses available provide an option for easier titration. The Freedom M [3] study utilized BID formulations. Additional ongoing studies are investigating oral treprostinil use with TID administration.
• Improvements in exercise capacity after 12 weeks of therapy.

Yet, several questions remain unanswered regarding the use of oral treprostinil in the treatment of PAH:

• What is the target dose?
  Patients in the Freedom M [3] study were not able to achieved their goal target dose due to tolerability. Newer lower dose formulations (0.25 mg) are available now, which theoretically should results in a higher maximal dose achieved.

• Is it effective on background therapy?
  FREEDOM-C2 [4] failed to meet its primary end point. PAH patients receiving background therapy (ERA, PDE-5I, or both) treated with oral treprostinil had a nonsignificant increase in 6MWD. The starting dose of oral treprostinil (1 mg BID) was later linked to poor tolerability and a relatively high rate of discontinuations because of adverse events.

• What do you do if your patient can not take oral medication and is hospitalized? What is the dose equivalent for IV and who should be able to prescribe oral medication?

• Does the oral formulation improve survival and long-term hemodynamics as have been demonstrated with IV or SQ formulations?
  Freedom M [3] was a short study that evaluated patients for 12 weeks and long term follow up studies are underway. Noteworthy, Betaprost, another oral prostacyclin approved only in Japan, showed improvement in 6 MWD at 3 and 6 months however the effect attenuated with time [5]. The compound never gained regulatory approval in the United States or Europe.

Orenitram is a very promising medication for the treatment of PAH. Stay tuned for additional studies evaluating efficacy and tolerability of this medication!

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• Consulting: Actelion, Gilead
• Speaker’s Bureau: Gilead, United Therapeutics

References:


Chronic Thromboembolic Pulmonary Hypertension (CTEPH) – Scan For It!

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One of the first patients I was assigned in the Internal Medicine residency clinic as an intern was a 23-year-old woman who had initially presented with worsening shortness of breath. Her work-up revealed multiple bilateral chronic pulmonary emboli and severe pulmonary hypertension. This was my first exposure to a patient with CTEPH – and I remember feeling quite overwhelmed when I realized how sick she was, and how little I knew of how to manage her! Under the guidance of my clinic preceptor, we referred her to the University of Michigan, our local tertiary care center. Through Vallerie McLaughlin’s detailed clinic notes, I learned more about this disease than I ever had read about in a medicine book. The patient was then referred to the University of San Diego for pulmonary endarterectomy but unfortunately was felt to be an inoperable candidate. I remember thinking, ‘so what else can we offer her’, only to realize that we had pretty much run out of options. She developed progressive right ventricular failure in spite of pulmonary vasodilator therapy and died before the age of 25. This incident left me wondering if I would ever get to see this terrible disease being conquered. And ten years later, having witnessed the exciting new developments in the CTEPH management, I can confidently say that we are getting very close! But the key to successfully managing these patients remains in early diagnosis and timely referral to experienced CTEPH centers.

CTEPH is a progressive disease which develops when thromboembolic material obstructs pulmonary artery branches causing a rise in pulmonary arterial pressure. It is quite rare but likely under-diagnosed, with an estimated 2-year incidence of 3.8% after first-time PE. However up to 60% of CTEPH patients have no history of PE. Whereas an abnormal echocardiogram usually triggers further evaluation, VQ scan (and not CTA) remains the preferred test for screening for CTEPH. Current guidelines suggest that patients with acute PE showing signs of PH or RV dysfunction at any time during their hospital stay should receive a follow-up echocardiography after discharge (usually after 3–6 months) to determine whether or not PH has resolved. A hemodynamic assessment with right heart catheterization is critical along with a high quality pulmonary angiogram to confirm the anatomy and location of clot burden. Once confirmed, patients should be assessed for pulmonary endarterectomy operability. There is an ever-increasing role of PAH targeted medical therapy in CTEPH management, particularly in those considered inoperable or with residual PH post operatively (but not as a replacement for PEA). Riociguat is an oral soluble guanylate cyclase stimulator that was recently approved for inoperable CTEPH patients.

The European CTEPH registry data has helped guide the field of CTEPH forward and a US registry is in the process of being established. The forthcoming ISHLT meeting in San Diego (April 2014) and
the International CTEPH Association conference in Paris (June 2014) will provide more/ new information in this exciting time of advancements in challenging this disease.

So, whenever I get the opportunity to diagnose and manage a CTEPH patient, I think back 10 years ago and feel more empowered with the tools and knowledge we have to tackle this disease now, instead of feeling overwhelmed by the grim prognosis.

Disclosure statement: The author has no disclosures to report.
Being a Physician Assistant in the Pulmonary Hypertension Arena

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It was about 3 years ago that I took the position of being a heart failure/pulmonary hypertension Physician Assistant (PA) at Allegheny General Hospital, Pittsburgh, PA. Although the time at AGH has been short, it has been filled with remarkable experiences. Our team specializes in advanced heart failure, pulmonary hypertension, mechanical circulatory support and cardiac transplantation. One of the areas in particular, pulmonary arterial hypertension (PAH), has always been fascinating. PAH is a relatively rare disease process, which has seen an explosion in terms of changes in understanding pathophysiology and treatment in the last decade. Three new medications were approved for the treatment of PAH this last year and many more novel therapies are available to our patients via clinical research trials. Being a part of this rapid and progressive change in medicine is thrilling as it is literally happening in front of our eyes!

The program would not succeed without a team-based approach to PAH patients. PAs serve as liaisons between the many facets of our group, working very closely with world-renowned physicians and surgeons, as well as dedicated PH pharmacists and nurse coordinators. One of the most important aspects of our team includes constant communication amongst one another, who have dedicated our lives to this disease and our patients. In rare diseases like PAH, communication is vital, and PAs guide patients’ transition from inpatient to outpatient by relaying detailed information to the nurse coordinators and partaking in direct patient care. It is a large responsibility for us to educate patients regarding their disease state and medications. In particular, when a PAH patient is newly started on a prostacyclin, there is a lot of counseling that is involved with an admission and discharge. PAs are also integrated into several clinical research projects that will hopefully shape the future of PAH treatment. The role of the PA includes screening and patient recruitment, communicating with research coordinators and physicians, and representing the institution at national PAH and related conferences like the upcoming ISHLT meeting in San Diego in April 2014.

Management of PAH is difficult as patients may seem well but are truly quite ill. Juxtaposed against the awe of PAH, is a truly humbling experience. I have sat with our patients and listened to stories about how their diagnosis has irrefutably changed their lives. Their gratitude for anything that can be done to extend and improve their lives is immeasurable. Bearing witness to their struggles only encourages us to work harder toward advancements in this disease. I feel honored to be a part of a team and the ISHLT that give so much to what is in the best interest of
patients with PAH. Being a PAH Physician Assistant cannot be summed up in the words above, but the best description for this career is exceedingly and indescribably rewarding.

Disclosure statement: the author has no disclosures.
**Next Frontier: Ventricular Assist Devices in Children With Failing Single Ventrices**

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We were honest with the parents and used terms such as “pioneering” and “innovative” to describe the operation we were offering. But we also admitted, “This surgery is still experimental. We do not know if it will work or not.” Our surgeon described how he would attempt to place DG, a 1 year old with a failing single ventricle, palliated to a bidirectional cavopulmonary anastomosis, onto a ventricular assist device (VAD). The procedure would involve an extensive reconstruction of the SVC and pulmonary artery, placing a modified shunt, followed by cannulation and connection to a rotary pump. He explained the risk of anesthesia and cardiac bypass. But as for the likely success of supporting a child with a failing single ventricle with a VAD? That was much more difficult to explain and quantify.

Over a 25-year period, from 1988 – 2013, there have been 16 published cases of children with single ventricular physiology supported with VADs (Table 1). Most cases involved pulsatile devices, a variety of surgical palliations, and support time of 2 to 363 days. Of these 16 reported cases, there was an overall survival of 63% (10/16). Encouraging, yet one must speculate about the number of similar unpublished cases whom were likely unsuccessful.

Weinstein and investigators examined the Berlin EXCOR Pediatric Investigational Device Exemption study database and identified 26 children with single ventricle anatomy implanted with the Berlin EXCOR [1]. Eleven of the 26 (42%) were successfully bridged to transplant and 11 died on EXCOR support. The remaining 4 were transitioned to ECMO due to failure of the VAD to provide adequate support and all four died within 30 days. In comparison, 185 of 255 (72%) biventricular patients supported with the EXCOR were bridged to transplant or recovery during the same time period.

Ten years ago, in 2004, our institution performed one of the first pediatric Berlin EXCOR device implantations in the United States in MC, a 5-month-old with cardiomyopathy. The device had been used in Europe, however the surgeons at our institution had yet to perform this procedure in such a small infant, nor did the medical team previously manage an infant VAD. The device, ordered from Germany, arrived with labels and manuals … printed in German. Our conversation back then with MC’s parents was probably not too different from our recent conversation with DG’s parents. **We are offering a pioneering, innovative therapy for your child. We do not have much experience with this. The outcome is uncertain, and he could die or suffer a devastating complication. Would you like us to proceed?**
According to the 2013 ISHLT annual report, 20% of pediatric heart transplant recipients were bridged with VADs or total artificial hearts [2]. Today, we can tell families that 75% of children on VADs are successfully bridged to transplant or recovery [3] and that postoperative survival is comparable to overall pediatric heart transplant survival [4]. Ten years ago, when we first started implanting VADs in children, we did not have these assurances.

Today, we are at a similar threshold, pushing the current boundaries with new applications of VAD to support children with univentricular heart disease. The challenges related to VAD support for this population are unique due to the complexity of single ventricle physiology and heterogeneous anatomy. VADs were well established in adults with biventricular circulation prior to application to children. In contrast, there are no larger population studies for single ventricle mechanical circulatory support.

Our current success in pediatric VAD support is due in part to families willing to accept the risks of a novel therapy and to medical teams willing to offer pioneering and innovative technology. The distinction between research and innovation may be challenging at these times, particularly without the infrastructure of a clinical trial. It is therefore imperative that we continue to share our successes and, perhaps more importantly, our failures with VAD support for children with single ventricle circulations, to inform the way.

### TABLE 1. Case reports of pediatric single ventricle patients supported with VAD

<table>
<thead>
<tr>
<th>Author</th>
<th>Year</th>
<th>Age (yrs)</th>
<th>Anatomy</th>
<th>Surgery</th>
<th>Device</th>
<th>Duration (days)</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Lal (5)</td>
<td>2013</td>
<td>1.2</td>
<td>HLHS (MS/AA)</td>
<td>Glenn-&gt;mBT shunt</td>
<td>Berlin then Revolution pump</td>
<td>65</td>
<td>OHT</td>
</tr>
<tr>
<td>Brancaccio (6)</td>
<td>2013</td>
<td>2</td>
<td>HLHS (MA/AA)</td>
<td>Glenn</td>
<td>Berlin</td>
<td>2</td>
<td>OHT (sudden death at 6mo)</td>
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<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
<td>Died (thromboembolic complications)</td>
</tr>
<tr>
<td>Mackling (7)</td>
<td>2012</td>
<td>4</td>
<td>DIRV</td>
<td>Glenn</td>
<td>Berlin</td>
<td>166</td>
<td>Died (sepsis)</td>
</tr>
<tr>
<td></td>
<td></td>
<td></td>
<td>DORV</td>
<td>Fontan</td>
<td>Berlin</td>
<td>363</td>
<td>Died (resp failure)</td>
</tr>
<tr>
<td>VanderPluym (8)</td>
<td>2011</td>
<td>3</td>
<td>HLHS (MA, TGA)</td>
<td>Fontan-&gt;Glenn</td>
<td>Berlin</td>
<td>174</td>
<td>OHT</td>
</tr>
<tr>
<td>Pearce (9)</td>
<td>2009</td>
<td>1.3</td>
<td>DORV/Mal-TGA</td>
<td>Central shunt</td>
<td>Berlin</td>
<td>49</td>
<td>OHT</td>
</tr>
<tr>
<td>Cardarelli (10)</td>
<td>2009</td>
<td>1.5</td>
<td>HLHS</td>
<td>Fontan</td>
<td>Berlin</td>
<td>179</td>
<td>Decannulation</td>
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<tr>
<td>Irving (11)</td>
<td>2009</td>
<td>2.9</td>
<td>HLHS</td>
<td>Fontan</td>
<td>Berlin</td>
<td>7</td>
<td>OHT</td>
</tr>
<tr>
<td>Chu (12)</td>
<td>2007</td>
<td>4</td>
<td>HLHS</td>
<td>Fontan</td>
<td>Berlin</td>
<td>13</td>
<td>Died</td>
</tr>
<tr>
<td>Calvaruso (13)</td>
<td>2007</td>
<td>10</td>
<td>Fontan</td>
<td>Berlin</td>
<td></td>
<td>7</td>
<td>OHT</td>
</tr>
<tr>
<td>Nathan (14)</td>
<td>2006</td>
<td>4</td>
<td>HLHS</td>
<td>Fontan</td>
<td>Berlin</td>
<td>28</td>
<td>OHT (died graft failure)</td>
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<td>Frazier* (15)</td>
<td>2005</td>
<td>14</td>
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<td>Fontan</td>
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<td>OHT</td>
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<td>Sadeghi (16)</td>
<td>2000</td>
<td>8</td>
<td>HLHS</td>
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<td>OHT</td>
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<td>Matsuda (17)</td>
<td>1988</td>
<td>10</td>
<td>Fontan</td>
<td>Toyobo (pulsatile)</td>
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<td>Died</td>
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<td>BT shunt</td>
<td>Toyobo (pulsatile)</td>
<td>5</td>
<td>Died</td>
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* Also reported by Russo 2008 (18)

HLHS hypoplastic left heart syndrome; MS mitral stenosis; AA aortic atresia; MA mitral atresia; AS aortic stenosis; DIRV double inlet right ventricle; DORV double outlet right ventricle; TGA transposition of great arteries; TA tricuspid atresia; OHT orthotopic heart transplant
Disclosure statement: the authors have no disclosures.

References:

Challenges in Pediatric Thoracic Transplant

Marc G. Schecter, MD
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The last ten (10) months have been a whirlwind. A new city, a new hospital, starting a new pediatric lung transplant program and a new role within the ISHLT have all occurred during this time. Each of these things has an element of uncertainty associated with them. Will the new program be successful? What is the culture of the new institution? How will my family adjust to a new city? Where are the good restaurants? Though these uncertainties have been ever present on my mind, they pale in comparison to the issues that plague our patients and their families. Will I be accepted for transplant? When will I receive my new heart or my new lungs? When I get my new organs, will they work? Our patients and families ask these questions every day. They trust us to make the right decision for their children.

Before a patient can be considered for a thoracic transplant, they must undergo a thorough evaluation to determine if the patient is an appropriate candidate. Most of the time, the patient’s underlying condition is the least complicated factor in determining whether a patient is a good candidate for transplant. The existence of co-morbidities certainly impact the decision of whether or not to approve a patient for transplant, but it is the confounding factors that often make our decisions more difficult, especially in pediatrics where we not only have to assess the patient, but the family as well. While assessing both the patient and their family, the multidisciplinary team of transplant experts determine the “red flags” and how much weight to give each one. If there are too many “red flags”, transplant may not be an option. There is always the possibility that a good candidate may be passed up, because of the system utilized. How good are we at identifying what really matters? Do we really know which “red flags” are more important? Did we deny a patient transplant that may have had an ideal outcome?

When we have a less than ideal outcome, it is always important to ask why. One of the first questions we should ask ourselves is whether we properly identified the patient as a good transplant candidate. Did we miss something during the evaluation that could have predicted the outcome? Did the cute faced patient blind us to move forward with transplant even though there was an increased risk of a poor outcome because of co-morbidities, family dynamics, or other issues? Did we succumb to the aggressive “I will do anything for my child” parent? Are we concerned about the medical and/or legal consequences of refusing to offer transplant to a patient? Should transplant be an option for everyone, regardless of the potential for worse outcomes? Is the media influencing us?

This year there have been three (3) highly publicized controversies in the United States regarding pediatric thoracic organ transplantation. They all involved the criteria utilized to select and list children for thoracic transplant. Historically, each individual physician/ transplant center has been
in control of their selection criteria, with very few hard and fast rules or guidelines governing the selection of patients for transplant. Some centers accept patients for transplant that other centers have denied. For better or worse, these public cases have brought the difficult decisions that we, as transplant physicians, make every day into the public arena, at times involving the legal system and most recently involving the United States Secretary of Health and Human Services. With the recent surge of cases in the media, it is unclear what the future will hold with respect to patient selection for transplant.

While these three (3) cases made national news in the United States this year, I am sure there are more stories both locally and internationally about who is appropriate for transplant and the method by which organs are allocated to patients. How are we to address these questions publicly when there may not be consensus among the pediatric thoracic transplant community? More importantly, do we need better or more stringent guidelines to address these issues? We gather every year at the annual ISHLT meeting and discuss new scientific advances and the medical conditions that affect outcomes. However, we rarely discuss how we account for these grey areas we deal with every day in selecting transplant candidates. Should we be reactive to situations in the media or be proactive to avoid the negative spotlight being placed on pediatric thoracic transplant?

The pediatric council asked the Board of Directors to begin this discussion at this year’s annual meeting in San Diego. On Saturday afternoon, there will be a symposium entitled "Controversies in Listing Children for Thoracic Organ Transplant". We have assembled a great group of speakers to stimulate the conversation. We hope this conversation will be the catalyst for a consensus conference that will provide cohesive guidelines for the thoracic transplant community. I encourage everyone to attend this symposium (Saturday, April 12, 2014, 2:00 pm - 3:30 pm, in Gaslamp CD).

See you in San Diego.

Disclosure statement: The author has no disclosures.
I changed institutions last year for the first time in over a decade. This sort of change often brings about an inclination to reminisce about the past. In this case my need to reminisce was not self-imposed but rather at the suggestion of UNOS who required a list of all my previous transplant experience prior to approving me as my new program’s Transplant Medical Director. I didn’t remember my patient’s UNOS numbers but I had no problem recalling the names of all my transplant patients over the past 25 years. Although I sent UNOS a list of all my patients, it was the ones with the poor outcomes whose details I could most easily recall. It was then I realized my biggest lessons learned in transplant were from my patients.

My first patient as a new attending physician was JJ, a teenager with familial cardiomyopathy. His uncle had received one of the first heart transplants at the center where I did my cardiology fellowship. Like many early recipients, he did not survive long and consequently, JJ’s mother was very hesitant to consider listing JJ for transplant. With the enthusiasm of a newly minted cardiologist, I reassured JJ’s mother that we had learned a great deal in the preceding decade and that our outcomes were improving all the time. We listed JJ. In the era before mechanical circulatory support, we supported JJ with inotropes in the ICU while we waited, hoping he would remain stable enough to avoid ECMO. We were lucky to get a donor organ quickly and his immediate post-transplant course was uncomplicated. He returned to playing his beloved baseball. Nine months after transplant his mother called me to say he had abdominal pain. I told her to bring him to the hospital. Shortly after arriving in the emergency room, he arrested and was placed on the previously avoided ECMO. Things did not go well. He developed multi-organ failure and did not recover. I found myself at the bedside discussing withdrawal of support with his mother. I assured her that he would not suffer. She looked at me and said “Can we wait just a little while longer then since once he dies, my suffering will never end”. I went to JJ’s funeral and sat in the back. I watched his baseball glove being put in before the casket was closed. I left without talking to the family. I had told them that things would be better than they had been for his uncle. I was wrong. JJ taught me humility. He also taught me that when patients die, statistics and outcomes are meaningless to our families.

Another lesson came from VS, also a teenager. He received his heart transplant after presenting in cardiogenic shock and being placed on ECMO. His presentation did not allow time for us to get well-acquainted until after his transplant. Medically he appeared to be doing very well with no rejection and good cardiac function for over two years. Every time I saw him, he would say that he did not feel well with multiple vague complaints. I did many extensive evaluations and never found anything that would explain his fatigue and occasional aches. He assured me he was not depressed...
but said he had “just never felt well since the transplant”. He eventually presented in cardiogenic shock with acute rejection. He had stopped taking his medications. He did not survive. The week prior he had told multiple family members as well as his pediatrician “if the question is ever raised, I do not want to have another transplant”. In essence, he had committed suicide by non-compliance. I do not know why VS never felt well and I have never had a patient like him again. Before VS, I prided myself on listening attentively to my patients. VS showed me that although I am listening, I am not always hearing. VS taught me a good ear is sometimes more important than another test. I hope I have learned to hear these patients now.

The last patient’s case taught me never to stop learning. I took care of AM for 18 years after his first heart transplant. He did great without discernible coronary vasculopathy or significant rejection. He received a second transplant for non-specific graft failure at 19 years of age. Initially he did great with no cellular rejection and great graft function. By the time of AM’s second transplant, we had learned a lot more about transplant immunology and we closely looked for DSA and AMR. This testing, too, was negative so it seemed AM’s second transplant would be long lasting. He died 11 months post-transplant with severe diffuse coronary vasculopathy. I had cared for AM for over 19 years. For a pediatric transplant physician that means multiple visits per year, multiple inpatient admissions and multiple soul-searching conversations with the family. I had watched him grow into a talented young man heading off to art school. I had celebrated his achievements and even his first detention (proof that he was living a “normal” life). When he was not telling very bad (actually, awful) jokes, he shared his plans, hopes and dreams. I so wanted to secure that future for him. I cannot tell you what led to his diffuse vasculopathy but I have not stopped looking. I continue to test his serum as we learn about new immune related factors that might explain rapidly progressive vasculopathy in a patient with none of the usual risk factors. I watched him grow over 19 years and then watched helplessly as he died. I did not sneak silently out of the back of his funeral. Surprisingly and at the family’s request, I spoke on his behalf … I guess we had both grown up a little.

Whenever I start feeling cocky and think I know so much about transplants, I think of JJ, VS and AM. I thank them for their lessons of humility, attentiveness and inquisitiveness. Despite our progress, we still have a lot to learn. It is what I love about transplant medicine … and occasionally it is what I hate about it. Reminiscing makes me realize we have come a long way in pediatric thoracic transplant, but when we measure success by decades rather than years, we still have a long way to go.

I am excited about the future of transplant medicine and plan on continuing to learn as much as possible from every patient.

Disclosure statement: the author has no conflicts of interest to report.
2014 Recipients of the ISHLT Leach-Abramson-Imhoff Links Travel Awards

The ISHLT Leach-Abramson-Imhoff Links Travel Awards, funded in part by the generous support from W.O. and Joan Leach (Gadsden, Alabama, USA), Mrs. Sue Abramson (Birmingham, Alabama, USA) and Mr. Larry Imhoff (La Place, Louisiana, USA), were created to support the growth and development of our future leaders from within our society including physicians, nurses, and other health care professionals. Those motivated enough with investigation, communication, and dissemination of new ideas for the betterment of patients with failing lungs and/or a failing heart including such conditions as pulmonary fibrosis, cystic fibrosis, emphysema, pulmonary hypertension, and from ischemic, nonischemic to congenital heart diseases should be awarded for their efforts.

Eligibility requirements include:

1. Any healthcare professional including but not limited to nurses, nurse coordinators, social workers, pharmacists, therapists, dietitians and early career physicians are eligible and must be a member of the ISHLT regardless of duration in their career.
2. An imposed restriction on physicians is that they must be in their Early Career—within 7 years of training, Assistant Professor equivalent, or junior faculty level with rare exceptions.
3. Individuals must display some form of research interest, basic, clinical, translational or outcomes investigations or at a minimum display some skill in journalism best exemplified by their contributions to the Links Newsletter engendering fresh and creative ideas.

Each year, the winners are selected from a pool of nominees by the ISHLT Links Travel Award Committee (LTAC). This committee includes the following individuals: the Links Editor-in-Chief, ISHLT Executive Director, ISHLT President, ISHLT Program Chair, and the Links Managing Editor.

The 2014 winners of the ISHLT Leach-Abramson-Imhoff Links Travel Awards were a cut above the other candidates for this year’s writer awards. With the rapid movement of replacement therapy, the importance of understanding the immune system, transplantation and prevention cannot be emphasized enough. Let’s extend a warm ISHLT congratulation to these writers.

**Writer of the Year: $2,500**

Christopher R. Ensor, PharmD, BCPS-CV  
Assistant Professor, Pharmacy and Therapeutics  
Clinical Faculty, Thoracic Transplantation  
University of Pittsburgh Medical Center  
Pittsburgh, Pennsylvania, USA
Our former-Chair and founding member of the Pharmacy and Pharmacology Council, Christopher Ensor, delivered an outstanding “Year in Review” (May 2013). But it was his clear, succinct and well-orchestrated summary on “Pneumocystis Jirovecii Pneumonia Prophylaxis After Lung Transplantation: The Bactrim Story…” (Dec 2013) which was nothing short of spectacular setting him apart from the rest earning this year’s Writer of the Year. His creative approach and assemblage about a very simple and inexpensive preventive strategy structured with 1) the challenge, 2) the data, and 3) the conclusion, has allowed us to eliminate the Pneumocystis jirovecii pneumonia in lung transplant recipients.

First Runner-Up: $1,500

Amanda Ingemi, PharmD
Transplant Clinical Specialist
Sentara Norfolk General Hospital
Norfolk, VA, USA

In keeping with the theme on prevention, another Pharm-D writer provided an important article on “Vaccines: the First Line of Infection Prophylaxis” (Sept 2013). Amanda Ingemi of Norfolk, Virginia is this year’s first runner-up. This clear, concise and organized article gives us an important strategy with a checklist of the appropriate vaccines necessary for successful transplant outcomes.

Honorable Mention: $1,000

Simon Urschel, MD
Clinical Director Pediatric Cardiac Transplant,
Assistant Professor of Pediatrics and Immunology
University of Alberta / Stollery Children's Hospital
Edmonton, AB, Canada

Simon Urschel’s article, First Naive, Later Delinquent: How Immune Maturation Benefits and Jeopardizes Transplantation (March 2013), shares with us the duality of the reality that children are different. The sickest among the sickest infants have the best conditional survival rates after heart transplantation. His article summarizes today’s understanding of the natural maturation of the immunization from infancy to independence right into adulthood.
External Grant (Yale, USA) Supports UMCU Research Collaboration

Although heart transplantation is a lifesaving procedure for end-stage heart failure patients, it is still challenged by rejection processes. In the quest of fighting rejection, microRNAs are suggested as possible key factors.

For this research project, Manon Huibers (PhD student under supervision of Roel de Weger, Pathology UMC Utrecht, NL) visited the lab of professor George Tellides at Yale School of Medicine (New Haven, USA) last year. This 4-month visit was amongst others supported by a travel grant of the International Society of Heart and Lung Transplantation (ISHLT).

This successful collaboration is now followed up by practical work in both labs (Tellides-lab USA and de Weger-lab NL). The Department of Surgery Ohse Research Grant Program at Yale School of Medicine awarded this collaboration with a $40,000 grant. The funds will be used in the coming year (until summer 2015) to support expenses for this collaborative project at Yale School of medicine.
Pediatric Transplantation Council Opportunities

**Associate Editor for Pediatrics**
**ISHLT Links Newsletter**
Seeking any eligible pediatric council member and Links enthusiast. We have an exciting opportunity for a council member to get involved with the ISHLT News Link as the Associate Editor for Pediatrics. A big thank you to Christian Benden who represented the Pediatric Council as the Associate Editor from 2012-2014.

**PEDS Council Vice Chair**
It is time to submit nominations for Vice Chair of the ISHLT Pediatric Council. Elections for the 2014-2015 Vice Chair will be held online after the ISHLT Pediatric Council Meeting in San Diego April 2014.

If you are interested in either position, please submit a statement to Marc Schecter (Marc.Schecter@cchmc.org) or Janet Scheel (JScheel@childrensnational.org) by **April 1, 2014**. Please contact any of the current Pediatric Council Leaders with questions.

These are great ways to get involved!
Outta This World Links
Interesting, Inspiring and Intriguing Links from Around the Globe

FROM AUSTRALIA:

Organ donations can save lives
Lung recipient Melissa Graham will be forever grateful
Melissa Graham knows she would not be alive today if it had not been for a generous donor who gave her new lungs. She was in her second year of studies at the University of New England, and she says "when you are 22 you think you are kind of invincible." But Melissa developed a condition called primary pulmonary hypertension, and needed a double lung transplant. Luck seems to have been on Melissa's side, within four weeks a donor was found, and nine years later she is still able to appreciate the gift she has received. Read more:
http://www.abc.net.au/local/stories/2014/02/27/3953294.htm?site=newengland

FROM IRELAND:

Joy at 'miracle' double lung transplant for Castlebar man
A young cystic fibrosis sufferer from Castlebar, who was staring death in the face last week, is recovering in hospital after a successful double lung transplant operation. Tom Concannon (25) from Turlough Road, Castlebar, was losing his battle with CF, a genetic disorder that affects the lungs and digestive system and dramatically reduces individuals’ life expectancy. However his mother Sandra said “a miracle” occurred on January 29 when a call came at 11pm announcing donor organs had been found. An ambulance arrived within 10 minutes and Tom was rushed to the Mater Hospital, Dublin, for the double transplant operation. “It’s a miracle, an absolute miracle,” said Ms Concannon. “Tom was losing his battle. He was on the donor list for a year but he never lost hope, he never gave up. The operation has been a success and we are getting closer and closer to going home.” Read more:

FROM THE UNITED KINGDOM:

Mum marks transplant milestone
Miracle double-lung transplant mum Natalie Kerr is spending today on the beach - to mark two years since her life-saving transplant.
The 32-year-old from Adlington had a successful transplant after being diagnosed with pulmonary hypertension. Now two years on, Natalie is celebrating being here to see her two children grow up and live her life to the full. Natalie has made it her goal to raise awareness of the transplant register and get more people to sign up. To mark the special milestone she has spent the past few
days in Lanzarote with her children Brandon, 11, and Isabelle, six, and other family members. She said: “It’s a really big day. It is two years since I got my new lungs, two extra years of living, two precious years with the kids. Read more: 
http://www.chorley-guardian.co.uk/news/local/mum-marks-transplant-milestone-1-6444862

FROM THE UNITED STATES:

Screenings and new treatments for heart defects can take patients from fetus to old age
Miami Herald Health
Tina Morton knew something wasn’t right in the 19th week of her pregnancy. Last year, Morton, 32, was sent by her obstetrician for a routine ultrasound. “We were excited to find out if we were having a girl or a boy,” she remembers thinking as the assistant rubbed the transducer across her belly. But then the assistant stopped, and the mood in the room changed. The technician called Morton’s doctor. “She couldn’t tell me anything, legally, but as a mom you know. I was listening to some of the terminology, and my husband looked at me and said, ‘What’s wrong?’ I said, ‘Something is wrong with her heart.’ ” Morton is a nurse. She knew she would need a field echo ultrasound to check the tiny baby’s heart. She wound up with a team of cardiac doctors at the Heart Institute of Joe DiMaggio Children’s Hospital in Hollywood. Daughter Addison, now 5 months old and thriving, was born with a malfunctioning left ventricle, and underwent open heart surgery when she was 10 days old. Today, advances in early detection and treatment for children with heart disease allows for more happy outcomes like Addison’s. Read more: 

Mesquite middle-schooler with heart diseases follows passion
The Dallas Morning News
When Carlee Baladez dances, she follows her heart. Carlee, a Mesquite Berry Middle School student, has already undergone two heart transplants in her 14 years. Carlee suffers from cardiomyopathy, a set of diseases that cause the heart to enlarge or thicken. She contracted the condition at age 1 after being exposed to chicken pox. But it has not stopped her from doing what she has always loved — dancing. “I want to be a professional ballerina — that is my goal,” Carlee said. ... The only sign Carlee showed of illness was the blisters on her body, but over time she showed symptoms of upper respiratory issues. “We just thought it was allergies, since she started showing symptoms around allergy season,” Berthelette said. Doctors at Children’s Medical Center in Dallas discovered her heart was enlarged to the point it was weighing on her lungs. After three months of medication and testing, Carlee once again came down with illness caused by a lowered immune system. Doctors determined Carlee needed a heart transplant. After a month’s worth of testing she was placed on the donor list. Read more: 
Parents Who Donated Their Child’s Heart Gave My Son the Gift of Life

The Stir

When Nicole Rogerson picked up the phone late one Tuesday evening in late January, she didn't know this would be the call that would change the life of her little boy. Nicole and Brian Rogerson’s son, Bryce, was born in 2005 with hypoplastic left heart syndrome (HLHS), a congenital heart defect in which the left side of a child’s heart is underdeveloped. Her miracle baby’s first heart surgery was in utero, when Nicole was just 26 1/2 weeks pregnant. His second took place at just 10 days old. But by the time Bryce was 6 years old, doctors told the Rogerson family that the heart they’d cut open and repaired time after time was putting too much stress on his lungs. The 6-year-old, they said, was going to need a heart transplant. That was March 2011, three years ago, when doctors listed Bryce on the transplant list. And then came the call on January 14. Doctors had found 8-year-old Bryce a new heart. Read more:
http://thestir.cafemom.com/big_kid/168227/parents_who_donated_their_childs

“Miracle Child” Receives Full Lung Transplant

CBS News

Her life story is nothing short of a miracle. Nancy Magana is one of just hundred babies who’ve received a full lung transplant at eight-weeks of age. She was born with a genetic mutation that causes the surfactant protein B deficiency. “It was the day we knew that she was sick” says her mother Fernanda. “She got trouble breathing.” Her parents knew what to look for because they had lost a child to the same mutation in 2002. Nancy’s only hope — a complete lung transplant. The hospital rushed Nancy to Texas Children’s Hospital in Houston – one of the few places in the country to offer lung transplants for infants. Dr. George Mallory Jr, the medical director of the lung transplant program at the hospital, was one of the doctors to see her. After seven long weeks they found a donor. It could not have come soon enough for Dr. Mallory. “This girl would not have lasted many more days if we hadn't gotten the lung offered when we got it.” Read more:
Tattling Links
ISHLT Members in the News

Two ISHLT members are worthy of our undivided attention and deserve the spotlight in this month’s Tattling Links page. We think you will agree!

Cardiologist Sharon Hunt to Receive Hewlett Award
Stanford University School of Medicine News
February 2014

After years of research, patient care, teaching, winning awards, mentoring trainees, and traveling all over the world, Sharon Hunt, MD, has done almost everything a transplant cardiologist could do. Added to her list of accomplishments will soon be the 2013 Hewlett Award. Walter Albion Hewlett was known as a physician of rare compassion and extraordinary skills. He was recognized for his outstanding contributions to patient care and medical science, much like Sharon Hunt. The award is designed as a recurring tribute to Hewlett, a professor and executive head of the Department of Medicine from 1916 to 1925. Being chosen as the Hewlett Award recipient was humbling, says Hunt, Professor of Cardiovascular Medicine. “I looked at the list of previous awardees, almost all of whom I knew, and there are some pretty amazing people. It’s also special because it is particular to Stanford, close to home, and personal.” Read more →

Sweet named Marriott Professor
Washington University in St. Louis Newsroom
February 4, 2014

Stuart C. Sweet, MD, PhD, a world leader in pediatric lung transplantation, has been named the W. McKim Marriott, MD, Professor of Pediatrics at Washington University School of Medicine in St. Louis. The professorship is supported by a dedicated endowment established by the St. Louis Children’s Hospital Foundation in partnership with St. Louis Children’s Hospital and the School of Medicine. It honors W. McKim Marriott, MD, who was a noted professor and chairman of pediatrics at the School of Medicine before serving as the school’s dean from 1923-36. Professor Emeritus James P. Keating, MD, was the inaugural W. McKim Marriott, MD, Professor of Pediatrics. Keating retired in 2012. “I am honored to hold this professorship, which reminds us of the important contributions Drs. Marriott and Keating made to Washington University and St. Louis Children’s Hospital,” Sweet said. “And I am grateful for the support provided by the St. Louis Children’s Hospital Foundation, which will help me continue our important work in pediatric transplantation.” Read more →
Editor’s Corner: Slang, Dang and Hang Me

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Jonathan Lighter, one of the leading authorities on American slang and editor of the Random House Historical Dictionary of American Slang, defines slang from Chapter Six of 2001’s Cambridge History of the English Language. He writes, “slang denotes an informal, nonstandard, nontechnical vocabulary composed chiefly of novel-sounding synonyms (and near synonyms) for standard words and phrases; it is often associated with youthful, raffish, or undignified persons and groups; and it conveys often striking connotations of impertinence or irreverence, especially for established attitudes and values within the prevailing culture.” Lighter further points out that “slang aims to be intentionally undignified, startling and amusing.” Think about when something “slips your mind” or when you “lose your train of thought.” Is it a “senior moment?” or a “brain fart?” The latter one challenges the standard, thus it is slang.

Slang comes from our younger and rebellious population. Perhaps it’s just part of being adolescent with a desire to be linguistically revolutionary outside the mainstream or against the establishment. How about a clever way to be critical of something we may be uncomfortable with such as change. Or a new concept or novel idea that was not originally our own. Slang needs to be novel, new or fresh and as such is typically ephemeral because it is revolutionary, unconventional and challenging.

In his book, Slang: The People’s Poetry, Michael Adams points out the association of slang with poetry, dating back to Ralph Waldo Emerson who celebrated slang as the poetry of language in action and that slang is our play with words, sort of a linguistic mischief with social and political statements as we poke fun. Slang is about the everyday; it’s not technical nanotechnology, immunology, fast-paced genomics, the internet or literary theory or linguistics, although all of these have their own jargon. Michael Adams adds to Lighter’s definition about the ‘insiderness’ of slang or the shared ‘outsiderness’, by stating, “slang meets our complementary needs to fit in and to stand out.” He also emphasizes that slang is “a necessary aspect of our linguistic well-being”; it’s not aberrant, deviant, or thoughtless, and it’s certainly not “unmeaning.” It is thoughtful and meaningful.

About over the same life span of the ISHLT, the ephemeral nature of some slang terms has morphed over time to “near-standard” acceptance. The word “bad” meaning good and “bummer” for an unpleasant experience are a couple examples. The word “cool” began nearly a century ago meaning sophisticated or up-to-date and lately meaning excellent. Our university students have many different ways to say good and cool as well as words they’ve
turned around from being bad to good as a playful example of slang. Other words for good include: bad, sick, wicked, killer, outrageous and now gnarly. There are also other meanings of good in general: sweet, candy, righteous including other words that have been elevated in intensity over time: awesome, choice, tight and rad. All of these slang examples are synonyms for good.

With the San Diego meeting upon us we might find our younger members beginning conversations with one another in some slangy manner with terms like: “Yo,” “What up?” “Sup?”; “What’s happening?” “What it is?” Many of us “old farts” might ask, “How are you?” will anyone ask, “How do you do?” or “What’s going on?” or a simple and easy “Hi” or “Hello.” Then to end conversations we might hear, “Later,” originating from “check you later” or “catch you later”; or “ciao,” “gotta go,” “gotta bounce,” and “I’m out.”

The playfulness of slang comes from creating new words or changing their meanings. With rhyme, cockney rhyming and literary alliteration the “boob tube” emerged defining our television or telly (Australian slang), meaning the idiot box. What about the terms “trouble and strife” – wife, “bacon and eggs” – legs, “apples and pears” – stairs, Adam and Eve – believe, “bread and honey” – money (note over time this has been shortened to bread also meaning money), Oxford scholar – dollar, and ducks and geese – police. Other rhyming slang terms are: bedhead, brain drain, fat cat, nit wit, no show, lovey dovey, tighty whities, chill pill, float your boat and balls to the wall.

Trying to understand slang or feel slangy could certainly make one feel queasy. You might recall “throw up” down under “in Australia” is “chunder.” In medicine there is emesis, in formal English there is vomit, and from our rebellious college students or colleagues we have many informal and slangy alternatives: barf, ralph, puke, spew, hurl, blow chunks, upchuck, deliver street pizza, drive the porcelain bus, pray to the porcelain gods, talk to ralph on the big white telephone, lose your lunch, blow chow and toss your cookies.

With so many words or terms for vomit, there are many more words or terms for a “collegial” cause of puking, that is none other than being drunk. The formal words are intoxicated and inebriated. The more slangy words or terms include: smashed, trashed, wasted, sloshed, loaded, soused, sauced, plastered, pissed, plowed, hammered, soused, plastered, bombed, blitzed, boozed, blotto, blackout, fried, stewed, stoned, sloppy, juiced, lit, pickled, tight, muddled, faded, loose, chunk, zooted, shwasted, hammed, at ham city, three sheets to the wind, tied one on, under the table, and I refer you to Benjamin Franklin’s published 200 terms for drunk in the Pennsylvania Gazette in 1737 with just these few for example: addled, bowz’d, cracked, wamble crop’d, he’s in his cups, he’s loaded his cart, he had a kick in the guts, he’s loose in the hilts, he has swallow’d a tavern token, and he’s got his top gallant sails out. I believe it’s time to be a bit abstemious.

I could on and on with this, but I refer you to the internet to pursue and peruse the use of slang in our everyday language and in the ISHLT. Finally, be sure to look up the Grammy
Award winner for Best Country and Western Song from half a century ago in 1964 by American country music artist Roger Miller with his song “Dang Me.”

To those of you who missed an extraordinary Oscars, I give you a taste with extraordinary songs, first by an extraordinary Irish group (U2) about an extraordinary man, Nelson Mandela, and freedom (Ordinary Love). The other songs that touch hearts and stir souls are about inspiration by Bette Midler (Wind Beneath My Wings), about destiny by Idina Menzel (Let it Go), and of course with the Academy's tribute to the 75th Anniversary of Judy Garland and the Wizard of Oz where the dreams you dare to dream really come true as we expand our minds (Somewhere Over the Rainbow).

Disclosure statement: The author has no conflicts of interest to disclose.
Word of the Month:

**Peripatetic** – related to Aristotle who taught and conducted discussions while strolling around the Lyceum. Today, it means to walk or move about, traveling from place to place; itinerant. And you might recall the line from the finale, “One” from the musical hit, A Chorus Line. One, singular sensation every little step he takes...she’s uncommonly rare, very unique, peripatetic, poetic and chic...

Word of the Month (slang):

**Lace up** – popularized by Cleveland, Ohio’s rapper, Machine Gun Kelly (Richard Colson Baker) meaning... as the word implies, lace up your kicks and do whatever it is you have to do, step up your game and take whatever life throws at you. Life is hard but you just should get ready and wait for what life brings you and don’t give a damn about what stress you have at the moment because destiny will make it.

Quote of the Month:

“I can always guess how many jellybeans are in a jar, even if I am wrong.” – Brick Tamland

You stay classy, San Di-Ahh-go