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The Optimized Parameters of Red Blood Cell Exchange by Apheresis in Transfusion-Dependent Thalassemia, a Small Case Series

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The Optimized Parameters of Red Blood Cell Exchange by Apheresis in Transfusion-Dependent Thalassemia, a Small Case Series

Abstract

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livedo racemosa on the lower abdomen, and lower back with mottled violaceous reticular patches plus punctuate ulcers. Skin biopsy revealed microvascular thrombi with no evidence of vasculitis. Laboratory work up was negative for hypercoagulable state. Immunofixation electrophoresis was positive for IgG kappa monoclonal protein. Neurology evaluation showed severe chronic axonal sensorimotor neuropathy on electromyography. Sural nerve biopsy was negative for amyloid and revealed perivascular mononuclear inflammation, scarred vessels with occlusion and areas of recanalization.

The clinical and histologic findings are consistent with livedoid vasculopathy (LV). The patient noted prompt improvement of his cutaneous findings and gradual improvement of his sensorimotor axonal neuropathy, with 400mg of pentoxifylline three times a day, 325mg of aspirin daily, and 20mg of rivaroxaban daily.

**#94. Widespread Primary Nodular Cutaneous Amyloidosis Due to Local Plasmacytomas**

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*Mentor:* Erin X. Wei

**Program:** Dermatology

**Type:** Case Report

**Background:** Primary nodular cutaneous amyloidosis (PNCA) is a rare manifestation of amyloid deposition, presenting as waxy, yellow-to-brown nodules or plaques that can be asymptomatic, painful, pruritic, and can compress surrounding structures. A diagnosis of PCNA requires careful evaluation to rule out plasma cell dyscrasias and rheumatologic conditions.

**Case:** A 74-year-old female presented to the clinic with >10 firm, well-defined subcutaneous “cysts” on her head and neck for the past three years. An excisional skin biopsy revealed depositions of eosinophilic extracellular material with apple-green birefringence on Congo red stain, consistent with a diagnosis of nodular cutaneous amyloidosis. Differential diagnoses included primary nodular cutaneous amyloidosis (PNCA), primary systemic amyloidosis due to plasma cell dyscrasia, or systemic amyloidosis due to chronic inflammation or infection. Laboratory workup revealed slightly elevated IgG lambda at 1.65, ANA titer of 1:320, positive dsDNA, kappa > lambda free light chain elevations, and mass spectroscopy typed lambda AL amyloidosis. Bone marrow biopsy with immunophenotypic staining was nonrevealing, and no abnormal cells were seen on the peripheral smear to suggest dyscrasia.

**Conclusion:** LV is a chronic condition related to micro-thrombosis of dermal vessels, which can cause ischemia and ulcerations. In rare instances, LV can induce peripheral neuropathy with limited cases recorded in literature. Our case further supports the importance of recognizing vasculopathy, particularly LV, as a cause for peripheral neuropathy.

**#95. The Optimized Parameters of Red Blood Cell Exchange by Apheresis in Transfusion-Dependent Thalassemia, a Small Case Series**

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*Mentor:* Aleh Bobr

**Program:** Pathology

**Type:** Case Report

**Background:** Thalassemias are red blood cell disorders characterized by defects in globin chains, resulting in a microcytic hypochromic anemia. Severe variants require red blood cell transfusions as frequently as every 2-4 weeks. Since the body has no mechanism to intentionally eliminate iron, these patients also require chelation yet may still experience hemochromatosis. As an alternative, several centers have used red blood cell exchange (RBCX) in place of simple transfusions with various success. However, exchange parameters have yet to be defined for transfusion-dependent thalassemia (TDT), unlike for sickle cell disease (SCD).

**Cases:** We had 5 patients with TDT who underwent RBCX with the primary goals to stabilize iron overload and increase transfusion intervals while satisfying the Thalassemia International Federation goal hemoglobin of 9.5 g/dL. The RBCX parameters used are in Table 1.

<table>
<thead>
<tr>
<th>Parameter</th>
<th>TDT RBCX</th>
<th>SCD RBCX</th>
</tr>
</thead>
<tbody>
<tr>
<td>Transfusion Interval (weeks)</td>
<td>5</td>
<td>4-7</td>
</tr>
<tr>
<td>Pre-Transfusion Hematocrit</td>
<td>29 (Hb 9.5 g/dL or higher)</td>
<td>25-27</td>
</tr>
<tr>
<td>Post-Transfusion Hematocrit Target</td>
<td>37-38</td>
<td>30 in acute patients; 32-34 in chronic exchanges</td>
</tr>
<tr>
<td>Isovolumic Hemodilution</td>
<td>Commonly omitted to keep FCR at 30</td>
<td>Performed, if pre-transfusion hematocrit allows</td>
</tr>
<tr>
<td>Fraction of Cells Remaining (FCR)</td>
<td>30</td>
<td>30</td>
</tr>
</tbody>
</table>

**Conclusion:** RBCX was well tolerated with only infrequent hypotension, citrate side effects, access issues, and rare vaso-vagal reactions potentially associated with elevated hematocrits. The adjusted RBCX goals allowed for an increase from an average interval of 3 weeks between transfusions to 5 weeks between RBCX. Despite an increase in...
average blood utilization, ferritin was either stable or down trending.

**Conclusion:** With 188 procedures performed over 4 years we have demonstrated that RBCX with parameters specific to TDT can be performed safely and efficiently in TDT patients. To our knowledge this is the first report of TDT-specific RBCX parameters. Though blood utilization is higher with RBCX, it offers longer intervals between transfusions and stabilized iron overload, improving quality of life for patients.

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**#97. Quality Improvement: Reduce Trending of Serum Lipase in Pediatric Acute Pancreatitis Patients**

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3Department of Pediatrics, Division of Hospital Medicine, College of Medicine, University of Nebraska Medical Center, Omaha, NE, USA

**Mentors:** Dave Freestone, Andrew Huang, Lauren Maskin

**Program:** Pediatrics – Gastroenterology

**Type:** Original Research

**Background:** Acute pancreatitis (AP) is a leading cause of GI-related pediatric hospitalizations. At least two of the following are required for diagnosis: characteristic abdominal pain, elevated serum amylase/lipase three times the upper limit of normal, and imaging consistent with AP. We identified that too many children admitted with AP are having lipase levels trended regularly. A prime reason for this is often to determine clinical progression or disease severity. While serum lipase is a useful diagnostic factor, it is not prognostic. Baseline data at our affiliated children’s hospital from January 2021 to December 2022 showed that lipase was trended in 74% of AP patients. Notably, average length of hospital stay for patients who had lipase trended was about 2.5 days longer. Our quality improvement (QI) project aimed to reduce lipase trending to <10% by February 2024.

**Methods:** Our interventions involved provider education, electronic health record alerts when repeat lipases were ordered, and informational flyers posted in provider work areas.

**Results:** During the first Plan-Do-Study-Act (PDSA) cycle from February 2023 to July 2023, trending percentage reduced to 38%. We reviewed the barriers and repeated interventions, and ran a second PDSA cycle from August 2023 to February 2024. The latest results demonstrate a trending percentage of 45%.

**Conclusion:** Trending lipase can result in prolonged hospital stay, increased patient/parental anxiety, and increased cost burden, including additional testing that may be sought to explain rising levels. This QI project will extend more PDSA cycles in efforts to lower the unnecessary lipase trending in hospitalizations for AP.

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**#98. Is Medication Management in the First 28 Days of Life Associated With Acute Kidney Injury in Extremely Low Birthweight Neonates?**

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**Mentor:** Melissa Thoene

**Program:** Pediatrics - Neonatology

**Type:** Original Research

**Background:** Acute kidney injury (AKI) in extremely low birthweight (ELBW) infants is associated with increased morbidity and mortality. The reasons for these worsened health outcomes are not fully elucidated. Our group sought to evaluate associations between AKI during neonatal intensive care unit (NICU) hospitalization in ELBW infants and common pre- and post-natal medical treatments and laboratory values.

**Methods:** An IRB-approved retrospective chart review of ELBW infants (N=47) admitted to a level III NICU was completed. Data on AKI, medical interventions, and laboratory values was collected. AKI was defined as a rise in serum creatinine (sCr) of ≥ 0.3 mg/dL within 48 hours or SCr >1.6, >1.1, and >1.0 for infants corrected to <28, 28-29, and 30-32 weeks gestation, respectively. Logistic regression was used to analyze the association of AKI with diuretic, steroid, or antibiotic use; combined protein provision; maximum sCr, maximum sodium, or minimum sodium levels on day of life (DOL) 1-14 and 15-28; vasoressor use DOL 1-10; maternal antibiotic or NSAID use; and maternal hypertension.

**Results:** Infants who received vasopressors DOL 1-10; steroids DOL 15-28; or diuretics or steroids on DOL 1-14 had lower odds of AKI after adjusting for CRIB II score (Table 1). No other significant associations were found.

**Table 1. Factors Associated with Lower Risk of AKI in ELBW Neonates After Adjusting for CRIB II Scores.**

<table>
<thead>
<tr>
<th></th>
<th>OR (95% CI)</th>
<th>P value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Diuretic use DOL 1-14</td>
<td>0.19 (0.04-0.90)</td>
<td>0.04</td>
</tr>
<tr>
<td>Steroid use DOL 1-14</td>
<td>0.056 (0.006-0.517)</td>
<td>0.01</td>
</tr>
<tr>
<td>Steroid use DOL 15-28</td>
<td>0.128 (0.021-0.7931)</td>
<td>0.03</td>
</tr>
<tr>
<td>Vasopressor use DOL 1-10</td>
<td>0.121 (0.022-0.681)</td>
<td>0.02</td>
</tr>
</tbody>
</table>

**Conclusion:** These findings were unexpected. This could be secondary to prevention of volume overload, improved renal perfusion, or close monitoring of fluid status. We did not define AKI by oliguria, which could affect association between creatinine levels and steroid or diuretic use. Future research with a larger cohort is warranted.
#99. Right-Sided Horner’s Syndrome as a Complication of Vascular Ring Repair
Jaikaran Man Singh1, Thomas Blount2, Camille Hancock-Friesen3

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3Department of Surgery, Division of Cardiothoracic Surgery, College of Medicine, University of Nebraska Medical Center, Omaha, NE, USA

Mentor: Thomas Blount
Program: Pediatrics – Cardiology
Type: Case Report

Background: Horner’s Syndrome is a known complication of neck surgeries, including vascular ring repair. This most commonly occurs on the left side after repair of the right aortic arch with an aberrant left subclavian artery. There have not been many cases of right-sided Horner’s syndrome specifically recorded during the repair of right Kommerrell diverticulum with a left-sided aortic arch.

Case: A 10-month-old girl was found to have a left aortic arch with an aberrant right subclavian artery and esophageal indentation. She underwent elective repair of her vascular ring, with dissection of the aberrant right subclavian and suturing into the subclavian artery; followed by double ligation of the ligamentum arteriosum. The recurrent laryngeal nerve was noted and spared, and the patient was extubated in the operating room.

Post-operative course was complicated by high volume chylothorax, which improved after transitioning to a low-fat diet. The patient also developed ptosis of the right eye, which was initially attributed due to facial edema but did not improve after diuresis.

Conclusion: Right-sided Horner’s syndrome can be a post-operative complication of vascular ring repair.

#100. Two True and Unrelated: Inpatient Evaluation for Severe Thrombocytopenia
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Mentor: Alex Nester
Program: Internal Medicine & Pediatrics
Type: Case Report

Background: Understanding the etiology of thrombocytopenia in hospitalized patients is essential to effective management.

Thrombocytopenia in liver disease is due to increased consumption and decreased production. In patients undergoing low bleeding risk procedures, no platelet directed therapies should be given. If undergoing a high bleeding risk procedure and platelets less than 50,000, then thrombopoietin receptor agonists or platelet transfusion should be considered.

Case: A middle-aged male with hypertension, alcohol use disorder in remission, esophageal varices, and cirrhosis presented with mucosal bleeding. Initial platelet count was remarkably low at 1 x 10^3/uL. Esophagogastroduodenoscopy at admission showed esophageal varices without evidence of recent bleeding. He was empirically treated for infection, but no infection was identified. Ophthalmology evaluation demonstrated 2 mm ptosis on the right side with good levator function. The right pupil measured 5 mm in the dark and the left pupil measured 8 mm. The remainder of the ocular examination was normal. The patient met the diagnosis of Right Horner Syndrome.

The patient continued to clinically improve and was discharged home on twice daily diuretics. She had reduced ptosis on her follow-up appointment one month after discharge.

Conclusion: In thrombocytopenia due to liver disease, transfusions should be given prior to procedures with high bleeding risk. ITP is a less common diagnosis but should be considered in profound thrombocytopenia when patients do not respond to transfusions.

#101. Neonatal Pasteurella Multocida Meningitis
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3Department of Pediatrics, Division of Pediatric Emergency Medicine, College of Medicine, University of Nebraska Medical Center, Omaha, NE, USA
4Department of Pediatrics, Division of Pediatric Infectious Diseases, College of Medicine, University of Nebraska Medical Center, Omaha, NE, USA

Mentor: Andrea Green Hines
Program: Pediatrics
Type: Case Report

Background: Pasteurella multocida is a gram-negative coccobacillus that most commonly causes cellulitis after bites, scratches or licks from colonized cats, dogs or other domestic or wild animals. More serious and invasive infections can occur, especially in the elderly, immunocompromised and neonatal populations. We present a case of Pasteurella multocida meningitis in a 4-day old neonate.

Case: A 4-day old male was admitted to the hospital with a fever (102F) and irritability. Laboratory results were remarkable for a normal complete blood count and comprehensive metabolic panel and elevated procalcitonin (5.74 ng/mL). Blood, CSF, and urine cultures were obtained. Empiric ampicillin and gentamicin were initiated.
#105. Plantar Grasp Sign as a Screening Tool for Orthostatic Tremor (OT)

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2Dartmouth-Hitchcock Medical Center, One Medical Center Drive, Lebanon, NH, USA

**Mentor:** Erin Smith

**Program:** Neurological Sciences – Movement Disorders

**Type:** Original Research

**Background:** Orthostatic tremor (OT) is a rare neurological disorder characterized by a sensation of instability while standing. Very few clinical signs have been described for OT to date. Finding other symptoms and signs could prove valuable for this hard-to-recognize disease.

**Methods:** This protocol is part of the University of Nebraska Medical Center Orthostatic Tremor longitudinal study. It was noted that OT patients flex their toes and sometimes the foot arch while standing (Plantar Grasp). They reported doing this to “grab” the floor and improve stability. This paper analyses the diagnostic test characteristics of the patient’s self-reported Plantar Grasp, a new sign in OT.

**Results:** There were 34 OT patients (88% females) and 20 control patients (65% females). Eighty-eight percent of patients with OT reported the plantar grasp sign and none of the controls. The Plantar Grasp Sign was found to be very sensitive (88%) and extremely specific (100%) in our cohort. Non-weighted Negative Likelihood Ratio (NLR) was 0.12. And the 3% prevalence weighted NLR was so low that the negative post-test probability was close to zero (Table 1).

**Conclusion:** Due to its high sensitivity, specificity, and ideal likelihood ratio, we propose that the Plantar Grasp sign could be considered to screen patients with possible OT. Further studies are needed to determine the specificity of this sign in OT versus other balance disorders.

<table>
<thead>
<tr>
<th>Plantar Grasp Sign</th>
<th>EMG Positive (OT)</th>
<th>EMG Negative (No OT)</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>Positive Plantar Grasp Sign</td>
<td>TP (30)</td>
<td>FP (0)</td>
<td>30 PPV (100%)</td>
</tr>
<tr>
<td>Negative Plantar Grasp Sign</td>
<td>FN (04)</td>
<td>TN (20)</td>
<td>24 NPV (83%)</td>
</tr>
<tr>
<td>Total</td>
<td>34</td>
<td>20</td>
<td>54 NLR (0.12)</td>
</tr>
</tbody>
</table>

**Table 1. Plantar Grasp Sign Test Characteristics**

EMG- Electromyography, TP- True Positive, FN- False Negative, FP- False Positive, TN- True Negative, PPV- Positive Predictive Value, NPV- Negative Predictive Value, NLR- Negative Likelihood Ratio, PLR- Positive Likelihood Ratio

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#106. Application of the Karnofsky Performance Scale (KPS) in Inpatient Cancer Rehabilitation

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2Department of Physical Therapy, College of Allied Health Professions, University of Nebraska Medical Center, Omaha, NE, USA

**Mentor:** Samuel Bierner

**Program:** Physical Medicine & Rehabilitation

**Type:** Original Research

**Background:** The Karnofsky Performance Scale (KPS) is an assessment tool used in oncology to measure functional status. Functional outcome measures used in inpatient rehabilitation include functional independence measure (FIM) and GG score. Using KPS in acute inpatient rehabilitation may provide a consistent method of communication between the oncology and acute rehabilitation teams.

**Methods:** Ninety three patients were selected randomly from a database of cancer patients admitted to Madonna Rehabilitation Hospital between 7/1/2019 – 6/30/2021. Two independent observers determined the admission/discharge KPS score for each patient. Admission/discharge GG score was calculated in 63 patients and admission/discharge FIM score was calculated in 30 patients.

**Results:** Graphical analysis of length of stay (LOS) and delta KPS showed a positive correlation, but with significant scatter between 0 and 20 delta KPS score (Figure 1). Graphical analysis of LOS and delta GG score showed a positive correlation without significant scatter. For determination of initial KPS score, there was substantial agreement between each observer (kappa = 0.700). For the final KPS score at discharge, there was fair agreement (kappa = 0.304). For the difference between final and initial scores (delta KPS), there was slight agreement (kappa = 0.2). Logistic regression showed that the only significant predictor of return to acute care was the average delta KPS with an odds ratio (95% confidence interval) of 0.845 (0.764 - 0.934). Initial KPS score was not a significant predictor of return to acute care.

**Conclusion:** The KPS may not be the best tool for tracking functional status during inpatient rehabilitation.