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

Kristina Sevcik  
*University of Nebraska Medical Center*

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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 punctate ulcerations. 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.

**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.

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

Mitchell A. Taylor<sup>1\*</sup>, Trevor Lockard<sup>1,2\*</sup>, **Robert Borucki<sup>1</sup>**, Erin X. Wei<sup>1</sup>

<sup>1</sup>Department of Dermatology, College of Medicine, University of Nebraska Medical Center; Omaha, NE, USA

<sup>2</sup>College of Medicine, University of Nebraska Medical Center; Omaha, NE, USA

\*contributed equally

**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 non-revealing, and no abnormal cells were seen on the peripheral smear to suggest dyscrasia.

Positron emission tomography-computed tomography demonstrated diffuse lymphadenopathy of the head and neck, and excisional biopsy of cervical lymph nodes demonstrated few deposits of lambda-restricted plasma cells surrounded by diffuse amyloid deposition. A diagnosis of PNCA with local plasmacytomas was made, and the patient is scheduled to begin daratumumab therapy and intralesional triamcinolone for symptomatic lesions.

**Conclusion:** PCNA is a rare disease that can present with elusive and nonspecific findings. The potential link to systemic amyloidosis underscores the importance of thorough evaluation in cases of cutaneous involvement.

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

**Kristina Sevcik<sup>1</sup>**, Claire Jackson<sup>2</sup>, Shelly Williams<sup>1</sup>, Scott Koepsell<sup>1</sup>, Aleh Bobr<sup>1</sup>

<sup>1</sup>Department of Pathology, Microbiology & Immunology, College of Medicine, University of Nebraska Medical Center, Omaha, NE, USA

<sup>2</sup>Department of Pediatrics, College of Medicine, University of Nebraska Medical Center, Omaha, NE, USA

**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.** Overall, RBCX was well tolerated with only infrequent hypotension, citrate side effects, access issues, and rare vasovagal 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

**Table 1.** RBCX Parameters for TDT vs. SCD

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

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.

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

**Sandeep Puri**<sup>1</sup>, Amreen Masthan<sup>2</sup>, Dave Freestone<sup>1</sup>, Andrew Huang<sup>1</sup>, Lauren Maskin<sup>3</sup>

<sup>1</sup>Department of Pediatrics, Division of Gastroenterology, College of Medicine, University of Nebraska Medical Center, Omaha, NE, USA

<sup>2</sup>Department of Pediatrics, College of Medicine, University of Nebraska Medical Center, Omaha, NE, USA

<sup>3</sup>Department 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.

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

**Shannon Haines**<sup>1</sup>, Robin Humrich<sup>1</sup>, Elizabeth Lyden<sup>2</sup>, Nicholas Torbert<sup>2</sup>, Pramod Shrestha<sup>1</sup>, Ann Anderson Berry<sup>1</sup>, Melissa Thoene<sup>1</sup>

<sup>1</sup>Department of Pediatrics, College of Medicine, University of Nebraska Medical Center, Omaha, NE, USA

<sup>2</sup>Department of Biostatistics, College of Public Health, University of Nebraska Medical Center, Omaha, NE, USA

**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  $\geq 0.3$  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; vasopressor 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.

**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.

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

	OR (95% CI)	P value
Diuretic use DOL 1-14	0.19 (0.04-0.90)	0.04
Steroid use DOL 1-14	0.056 (0.006-0.517)	0.01
Steroid use DOL 15-28	0.128 (0.021-0.7931)	0.03
Vasopressor use DOL 1-10	0.121 (0.022-0.681)	0.02

## #99. Right-Sided Horner's Syndrome as a Complication of Vascular Ring Repair

**Jaikaran Man Singh**<sup>1</sup>, Thomas Blount<sup>2</sup>, Camille Hancock-Friesen<sup>3</sup>

<sup>1</sup>Department of Pediatrics, Division of Cardiovascular Medicine, College of Medicine, University of Nebraska Medical Center, Omaha, NE, USA

<sup>2</sup>Department of Pediatric Cardiology, Children's Nebraska, Omaha, NE, USA

<sup>3</sup>Department 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.

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:** 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

**Audrey Lane**<sup>1,2</sup>, Alex Nester<sup>3</sup>

<sup>1</sup>Department of Internal Medicine, College of Medicine, University of Nebraska Medical Center, Omaha, NE, USA

<sup>2</sup>Department of Pediatrics, College of Medicine, University of Nebraska Medical Center, Omaha, NE, USA

<sup>3</sup>Department of Internal Medicine, Division of Hematology & Oncology, College of Medicine, University of Nebraska Medical Center, Omaha, NE, USA

**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.

Immune Thrombocytopenia (ITP) is a diagnosis of exclusion. Patients are considered refractory to platelet transfusions when a 1-hour post-transfusion platelet count does not increase. Steroids and IVIG are first line treatments. Second line treatments include thrombopoietin receptor agonists and CD20 antagonists.

**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  $1 \times 10^3/\mu\text{L}$ . Esophagogastroduodenoscopy at admission showed esophageal varices without evidence of recent bleeding. He was empirically treated for infection, but no infection was identified.

Despite receiving multiple platelet transfusions ( $>20$ ), he continued to be profoundly thrombocytopenic (Corrected Count Increment of 0). He was for positive platelet antibody IgG and IgM. He did not respond to initial empiric treatments for ITP; specifically, four doses of IVIG, eltrombopag, and dexamethasone. Due to refractory nature, he received romiplostim followed by rituximab with platelet recovery.

**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

**Lauren Glasner**<sup>1</sup>, Lauren Maskin<sup>2</sup>, Hannah Sneller<sup>3</sup>, Andrea Green Hines<sup>4</sup>

<sup>1</sup>Department of Pediatrics, College of Medicine, University of Nebraska Medical Center, Omaha, NE, USA

<sup>2</sup>Department of Pediatrics, Division of Pediatric Hospital Medicine, College of Medicine, University of Nebraska Medical Center, Omaha, NE, USA

<sup>3</sup>Department of Pediatrics, Division of Pediatric Emergency Medicine, College of Medicine, University of Nebraska Medical Center, Omaha, NE, USA

<sup>4</sup>Department 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.

Blood and CSF cultures became positive for *Pasteurella multocida*. Parents reported a dog and cat in the home but denied direct contact with the neonate. However, the mother reported she sustained cat scratches and bites throughout her pregnancy, including the days up to delivery. Therefore, it is possible that the infection was incurred via vertical

transmission. Given the potential for suppurative complications of neonatal bacterial meningitis, a brain MRI was obtained and was normal. The patient completed 21 days of ampicillin and had a complete recovery. Consent was obtained to use this case for educational purposes.

**Conclusion:** Care should be taken with neonates exposed to household animals, as they are at risk for more serious and invasive *Pasteurella* infections. Notably, caution should also be taken with expecting mothers who are exposed to animals that are more likely to scratch or bite, as vertical transmission of *Pasteurella* is possible.

## #105. Plantar Grasp Sign as a Screening Tool for Orthostatic Tremor (OT)

**Vekash Raja**<sup>1</sup>, Rebecca Thompson<sup>1,2</sup>, Danish Bhatti<sup>1</sup>, Kalyan Malgireddy<sup>1</sup>, Venkata Sunil Bendi<sup>1</sup>, John M. Bertoni<sup>1</sup>, Diego Torres-Russotto<sup>1</sup>

<sup>1</sup>Department of Neurological Sciences, Division of Movement Disorders, College of Medicine, University of Nebraska Medical Center, Omaha, NE, USA

<sup>2</sup>Dartmouth-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 1.** Plantar Grasp Sign Test Characteristics

Plantar Grasp Sign	EMG Positive (OT)	EMG Negative (No OT)	Total	
Positive Plantar Grasp Sign	TP (30)	FP (0)	30	PPV (100%)
Negative Plantar Grasp Sign	FN (04)	TN (20)	24	NPV (83%)
Total	34	20	54	NLR (0.12)
	Sensitivity (88%)	Specificity (100%)	PLR: to infinity	Accuracy (92%)

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

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

**Justin Comer**<sup>1</sup>, Samuel Bierner<sup>1</sup>, Celeste Newstrom<sup>2</sup>

<sup>1</sup>Department of Physical Medicine & Rehabilitation, College of Medicine, University of Nebraska Medical Center, Omaha, NE, USA

<sup>2</sup>Department 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.