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PHYSICIAN REPORT

WINTER 2019

> REGINALD WASHINGTON, MD, FAAP, FACC, FAHA
Chief Medical Officer

Rocky Mountain Hospital for Children

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INSIDE

A Note from Dr. Washington

Welcome to the Winter 2019 issue of *Rocky Mountain Hospital for Children Physician Report*. In this issue, we invite you to learn how our physician teams worked together to develop a multidisciplinary plan to help heal a young patient suffering from chronic anemia.

Also, we're including information about innovative updates to several of our pediatric operating rooms. The upgrades support our focus on patient and physician satisfaction. See pages 2 and 3 for details.

While we thank all of our physicians for providing excellent care, we hope you join me in congratulating our 2019 Physician Spirit Award Winner. She was nominated by our nursing and clinical teams.

Finally, turn to page 4 to read an inspiring story about our genetics team and the importance of a Level IV NICU.

Sincerely,

Reginald Washington, MD, FAAP, FACC, FAHA
Chief Medical Officer

Teamwork at RMHC Helps Young Child Suffering From Chronic Anemia

A young child presented to Rocky Mountain Hospital for Children (RMHC) for the first time with recurrent syncope and was found to have a Hb of 2.9. History suggested chronic issues with anemia, necessitating transfusion. The cause of this anemia was not apparent.

> THEODORE STATHOS, MD
Rocky Mountain Hospital for Children

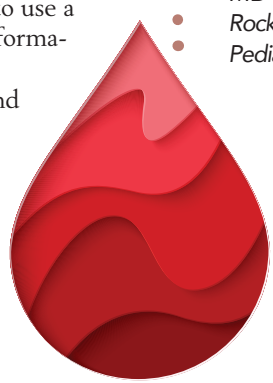
> SAUNDRA KAY, MD
Rocky Mountain Pediatric Surgery

A PLAN OF ACTION

The patient was admitted to the pediatric ICU where she received several units of blood. Work-up began and she was noted to have intermittent occult blood in the stool. The pediatric gastroenterology team led by Theodore Stathos, MD, was consulted.

An upper endoscopy revealed an arteriovenous malformation (AVM) in the duodenal bulb that was actively oozing blood. Cautery was used and several endoclips were deployed around the lesion to stop the bleeding, without the need for surgery. Dr. Stathos then deployed a pill cam to evaluate the rest of her small intestine for any additional lesions.

The RMHC GI team evaluated the images from the pill cam and noted at least five more AVMs in the small intestine, including a large one in the terminal ileum. Dr. Stathos then scheduled a repeat upper endoscopy planning to use a longer scope to try to reach the malformations in the proximal jejunum, and a colonoscopy to evaluate the colon and attempt to reach the lesions in the distal ileum. He was successful in cauterizing an AVM in the proximal jejunum and one in the terminal ileum. He was also able to perform a submucosal resection of a large polypoid AVM in the colon, allowing tissue for pathologic confirmation.



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OPERATING ROOM Updates Focus on Easing Patient (and Parent) Anxiety

Patient satisfaction is a highly regarded and a carefully monitored measurement of our patient-centered medical care. Three primary factors that have risen to the top of that measurement from the patient's perspective are:

- Good communication
- Personalized care
- Lowering anxiety of the patient and their immediate family.

To address the factors, Rocky Mountain Hospital for Children (RMHC) recently upgraded our dedicated pediatric operating rooms to create an improved patient surgical experience.

OPEN PATH TO COMMUNICATION

Highlights of the upgrade allow for a personal, calming introduction into the surgical suite for our pediatric patients. This introduction leverages technology and communication pathways to create an instant, easy way for the surgical team to connect with families during a procedure via secure text or e-mail. It is also available for use in delivering care and discharge instructions. This communication is especially helpful if patient families need to leave the immediate OR waiting area or hospital lobbies.

ADDING COMFORT FOR PATIENTS

A survey completed in 2018-2019 at RMHC answered by parents of children undergoing a surgical procedure indicated that 83% of respondents said that having their child watch a movie was effective in helping them remain calm and distracted during their visit. Further, in a recent clinical study, 99% of participants reported receiving text messages reduced their anxiety.

Aside from communication with parents/caregivers during surgery, RMHC has enhanced availability of video, music, serene landscapes and children's movies to lower patient anxiety and increase patient relaxation before anesthesia or during a procedure. The enhanced features include award-winning first-run movies and TV shows rotated quarterly.

Source: https://www.ncbi.nlm.nih.gov/pmc/articles/pmc1839580

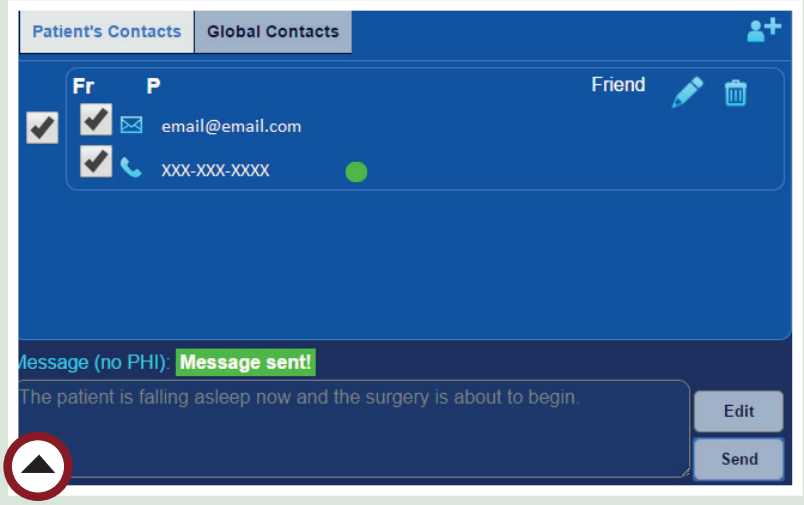

IMAGE 1.
The updated OR communication platform helps patients feel informed and calm. The updates are created easily in the operating room and delivered via mobile phone number or personal e-mail.

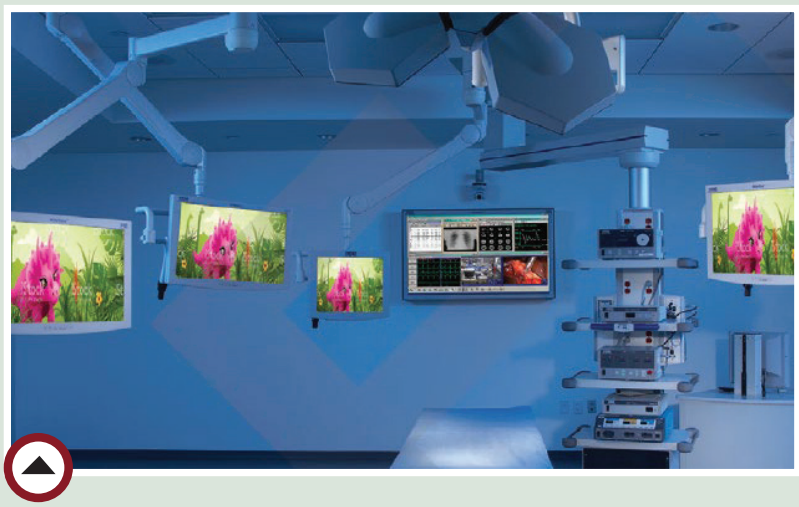

IMAGE 2.
A simulation of the enhanced entertainment options that greet pediatric surgery patients in our newer pediatric ORs.

For more information, contact Dana Zarcula, Director, Pediatric Surgical Services: **720-754-4266.**

RMHC Names Dr. Alice Liu 5th Annual Spirit Award Winner

Please join us in congratulating Alice Liu, MD, Pediatric Emergency Department Physician. Dr. Liu was nominated for the Annual Spirit Award by clinical teams at Rocky Mountain Hospital for Children. The award recognizes outstanding and consistent quality patient care, empathy, participation, a strong commitment to the hospital's mission and vision, exceptional support to local or regional communities and a strong connection to staff and colleagues.

"She makes every family feel welcome and eases their concern for their child, whether their child is having a life-threatening emergency or the family just needs support and reassurance… Her positivity inspires the entire team to continue to provide the best care we can for our patients and their families."

RMHC Patient Feedback Score Strengthens to 4.9 out of 5.0

In our last issue, we shared that our Patient Feedback Score was 4.7. We are thrilled to share that our score has now increased to 4.9 out of 5. In comparison, the competitive benchmark for our category is 4.4 out of 5.

Thank you to everyone who has had a part in our exceptional patient care.

(continued from page 1)

The patient had stabilized nicely and was discharged home with close follow-up. Unfortunately, the patient began bleeding once again and was readmitted to RMHC. Dr. Stathos performed an upper endoscopy to try to identify the bleeding source. The previously treated lesion in the duodenal bulb had involuted and was clearly not the source. A second pill cam was deployed and this suggested active bleeding from a distal ileal AVM. A repeat colonoscopy to try to control this was performed by Dr. Stathos. There was no active bleeding at the time but he did note a lesion at the ileocecal valve which was removed. The pediatric hematology team was consulted and the patient was started on a medication to try to shrink the AVMs.

There was a collective ongoing concern for this patient, as the child had multiple untreated AVMs throughout the small intestine. Thus, Dr. Stathos joined forces with Saundra Kay, MD, from Rocky Mountain Pediatric Surgery to come up with a treatment plan. They decided to perform an intraoperative enteroscopy, whereby Dr. Stathos's endoscope would be guided laparoscopically through the small intestine, allowing evaluation, and treatment of much more of the small intestine.

This uncommon procedure was scheduled with the two doctors for the next day. Dr. Kay began with three small laparoscopic incisions in the patient's abdomen and skillfully advanced the intestine over the GI scope so that Dr. Kay and Dr. Stathos could evaluate every inch of the intestine, Dr. Stathos from the inside and Dr. Kay from the outside. It became apparent that, in this case, all the lesions that Dr. Stathos spotted on the inside had an obvious abnormality noted on the external surface of the small intestine.

Dr. Kay continued her examination of the distal small intestine and noted multiple more AVMs, some large, others small. In the end, six AVMs were removed with multiple bowel resections and a wedge resection of a smaller lesion.

MULTIDISCIPLINARY TEAMWORK

Although the patient remains on medication to prevent enlargement of any tiny residual AVMs, the patient has been doing very well at home with a stable Hb and no signs of bleeding.

The exceptional teamwork at RMHC allows them to constantly think outside the box to provide the best care for all children.

Prenatal Presentation of Severe Familial Hypophosphatasia with Novel Mutation in ALPL Gene

> CATHERINE BURSON, MS, CGC
Senior Genetic Counselor

> BRIAN CORNER, MS
Genetic Counselor

> BRONWEN KAHN, MD
Maternal Fetal Specialist

> JAN KENNAUGH, MD.
Neonatologist

> SUNIL NAYAK, MD
Pediatric Endocrinologist

> JAREN RILEY, MD
Pediatric Orthopedics

> JOHN ROSS, MD
Neonatologist

The following is an abstract for a presentation delivered at the 2019 National Society of Genetic Counselors 38th Annual Conference by Catherine Burson, MS, CGC, Senior Genetic Counselor at Rocky Mountain Hospital for Children.

A prenatal ultrasound completed by Obstetrix Medical Group at their outreach clinic in Durango, Colorado, at 19 weeks indicated significant skeletal dysplasia, with severe mesomelia and angulated femurs. Further, chest circumference was at 10% with no identifiable rib fractures or beading. An amniocentesis was performed and in collaboration with the genetics team at Rocky Mountain Hospital for Children (RMHC), microarray and an OI/skeletal dysplasia panel was ordered.

The skeletal dysplasia panel came back positive for likely pathogenic mutation in ALPL gene. Mutations in this gene have been associated with both early onset recessive and later onset dominant hypophosphatasia (HPP) with poor phenotype-genotype correlation. Parental testing by the genetic team at RMHC confirmed that the mother carried the same mutation with no symptomology for HPP so the family was counseled that the diagnosis was still not confirmed and other possible diagnoses could be in the differential including campomelic dysplasia.

At time of delivery at the RMHC Level IV NICU, baby did well with minimal respiratory assistance. Alkaline phosphatase (ALP) levels were drawn and were significantly low. The low

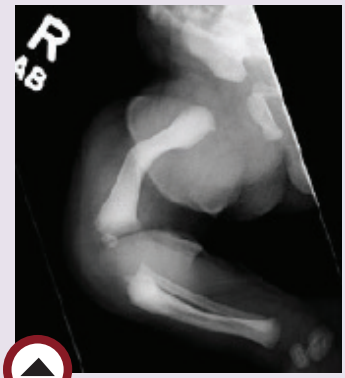

FIGURE 1.
X-ray at delivery

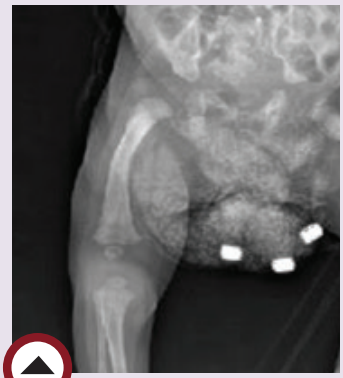

FIGURE 2.
X-ray at 6 weeks (5 weeks of treatment)

ALP results confirmed the diagnosis of hypophosphatasia; baby was started on enzyme replacement treatment at day seven of life with ongoing treatment and management by RMHC pediatric endocrinology and orthopedics. Subsequent ALP levels in mother and maternal grandfather were also low and family history was significant for muscle and joint pain and metatarsal break in maternal grandfather.

Familial variability is well documented in HPP; this case is particularly unique because of the seemingly discordant family history with absence of clinical signs in mother and severe prenatal presentation in baby. Incomplete penetrance of the dominant mutation is one possibility; familial variability with later onset in mother is another.

The more severe presentation could also be due to the combination of low maternal ALP levels in utero along with baby's underlying low ALP level. Lower levels during critical first trimester would have greater impact on bone development; maternal ALP levels are typically elevated during third trimester, which may have improved this infant's perinatal outcome.

Early enzyme replacement therapy has proven successful at preventing subsequent fractures in infants diagnosed prenatally with congenital HPP. Additionally, treatment has been shown to improve long bone bowing, with possibility of resolution and typical motor development. Other infants with described "benign" HPP have been identified. Although respiratory outcome may be better relative to lethal neonatal HPP, these patients still have significant skeletal sequelae and would benefit from treatment.

Follow-up at 6 months of age shows a healthy child with no new fractures identified. Additionally, long bone bowing is decreasing and baby's developmental milestones are on track. Reactions to the enzyme replacement therapy have been minimal.

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