

Shockingly Constipated: Severe Constipation and Multi System Organ Failure in a Child With Down Syndrome

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Background

- Constipation commonly occurs without severe complications, with prevalence of the disease up to 29.6% for children worldwide¹
- Those with developmental delays are at greater risk of severe complications, which can include rectal tears, rectal prolapse, pelvic dysynergia¹
 - Constipation risk factors for trisomy 21 also include hypothyroidism and Hirschsprung's Disease²
- Severe constipation is a risk factor for Intra-abdominal Hypertension (IAH) and Abdominal Compartment Syndrome (ACS)^{3,4}
 - IAH = sustained or repeated intra-abdominal pressure (IAP) > 10 mmHg
 - ACS = sustained elevation of IAP > 10 mmHg resulting in new or worsening organ dysfunction
- Treatment includes evacuation of intra-abdominal contents
- Studies suggest 40-60% mortality in pediatric ACS, even with early decompression⁵

Initial Presentation

- A 12-year-old male with trisomy 21 and chronic constipation presented with abdominal distension and vomiting
- He had not had a bowel movement for 5 days
- Mother reports that last Miralax was >2 weeks ago
- Initial vital signs were within normal limits
- Initial exam revealed mild abdominal distension with a firm rectal stool ball (rectal tone was normal)
- Baseline labs were unremarkable except for a hypokalemia (2.9 mmol/L) and slightly high creatinine at 0.8 mg/dL
- A CT scan showed fecal impaction resulting in bowel obstruction
- An NG tube was placed and GoLytely was administered per protocol
- Multiple enemas were given without bowel movement
- An IV was placed and maintenance fluids were started, though it was removed multiple times by patient
- Later that day, a nurse calls the physician to bedside due to "purple legs"

Exam

- The patient had an extremely distended abdomen with blanching of the abdominal wall, mottling of the thighs with thready femoral pulses, and duskiness of the feet with unappreciable distal pulses
- Mental status had also become altered
- Changes in his vital signs included HR in the 140s, BP 65/24, and O2 saturation 86%, consistent with shock

Images and Tables

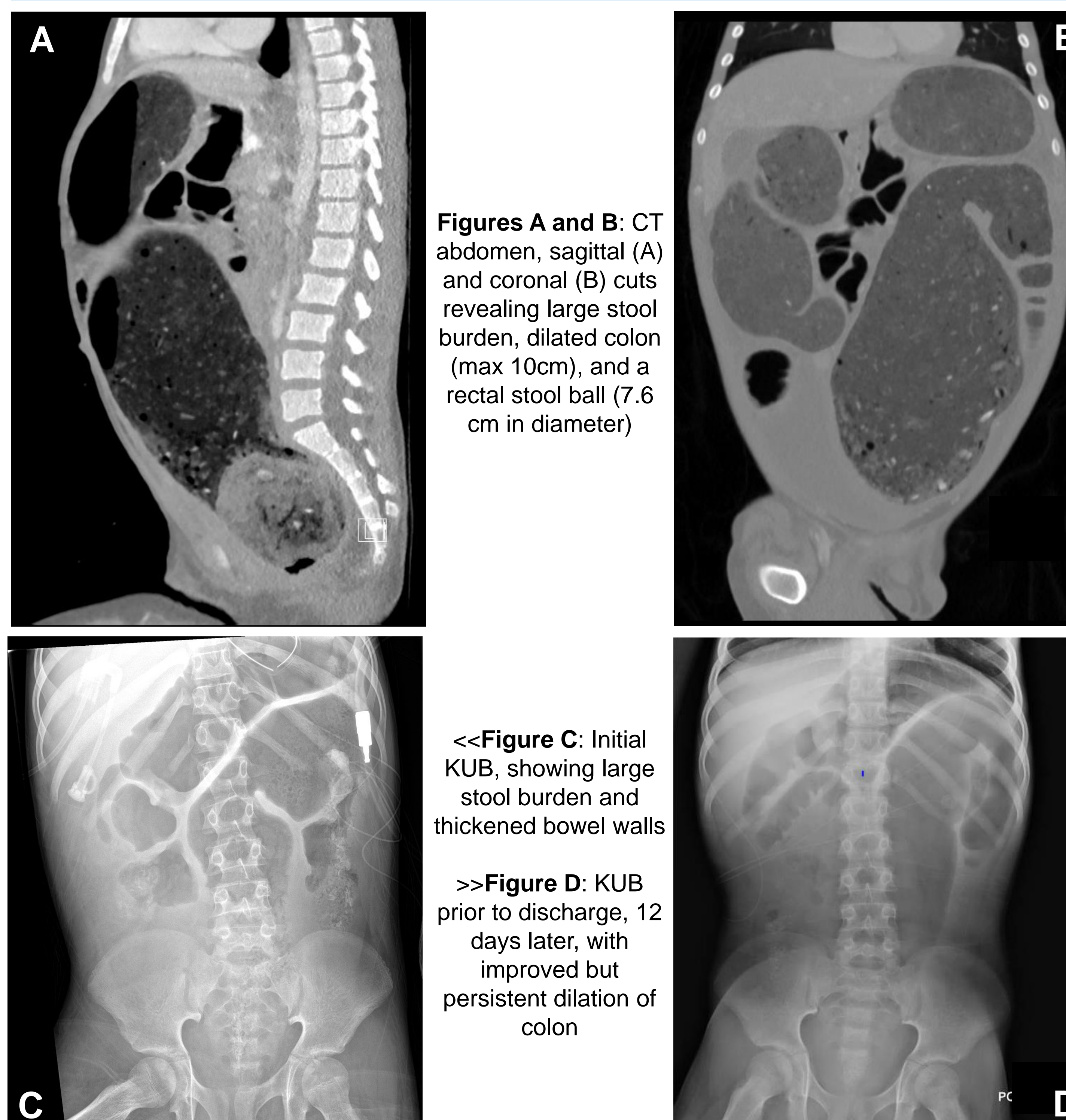


Table 1: Pertinent labs (*MSK labs reflect highest values during admission)

Table 1	Lab	At Admission	Repeat labs, 12hrs later	Ref. Range
Electrolytes	Na	135	140	135-145 mmol/L
	K	2.9 (L)	4.3	3.5-5.1 mmol/L
	Cl	99 (L)	107	101-111 mmol/L
	HCO3	20 (L)	12 (L)	22-32 mmol/L
	BUN	17	24 (H)	6-20 mg/dL
	Cr	0.83	1.7 (H)	0.5-1.0 mg/dL
	Gluc	129 (H)	430 (H)	65-99 mg/dL
	Albumin	4.1	3.9	3.5-5 g/dL
	AST	26	287 (H)	15-41 U/L
	ALT	18	317 (H)	17-63 U/L
	T bill	0.6	1.4 (H)	0.3-1.2 mg/dL
CBC	WBC	12.4	8.7	4.5-13.5 K/uL
	Hgb	12.5	13	11.0-14.6 g/dL
	Plt	522 (H)	475 (H)	150-400 K/uL
VBG	pH	-	7.03 (L)	7.25-7.43
	Lactate	-	14.2 (H)	0.5-1.9 mmol/L
Inflammatory	CRP	-	2.6 (H)	<1.0 mg/dL
MSK*	CK	-	8,453 (H)	49-397 U/L
	Myoglobin, Ur	-	391 (H)	0-13 ng/mL
Thyroid	TSH	-	2.76	0.4-5.0 uIU/mL
	Free T4	-	1.08	0.61-1.12 ng/dL

Clinical Course (cont'd)

- The patient was started on O2 and was emergently rectally disimpacted at bedside
- Copious amounts of liquid stool, including non-food items, were evacuated from the colon
- Resuscitation was simultaneously initiated with fluid boluses and pressors (dopamine, norepinephrine, and vasopressin)
- IAP was not measured due to the patient's critical condition
- Zosyn was started for presumed bacterial translocation and sepsis-like picture; blood cultures were negative
- Once vitals stable, patient was transferred to the OR for continued stool removal and subsequent ICU admission
- GoLytely was trialed again, though patient again became distended and required a second disimpaction
 - Further therapy consisted of Miralax and enemas only
- Patient remained intubated and ventilated for 5 days
- Course was complicated by acute tubular necrosis and consequent diuresis (max 32 mL/kg/hr), SIADH, rhabdomyolysis, acute liver injury, and *C difficile* colitis
- Patient was eventually discharged on hospital day 16 with a referral to GI at a subspecialty center to evaluate possible Hirschsprung's disease

Discussion

- Our patient experienced hypovolemic shock and likely ACS secondary to acute obstruction in the setting of chronic constipation
- Though IAP was not measured, patient did meet clinical criteria for ACS
 - Tense abdomen, decreased lower extremity pulses, and multisystem organ failure
- Hypovolemic shock likely due to a combination of 1] failure to maintain PIV access for hydration, 2] the osmotic load of GoLytely, and 3] decreased venous return
- Extra time counseling families on the importance of routine bowel regimens may be warranted for developmentally delayed children with constipation to prevent complications
 - Consider a more aggressive management approach (i.e.: NG decompression, manual disimpaction) than typical of routine constipation clean-out admissions for this population

References

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