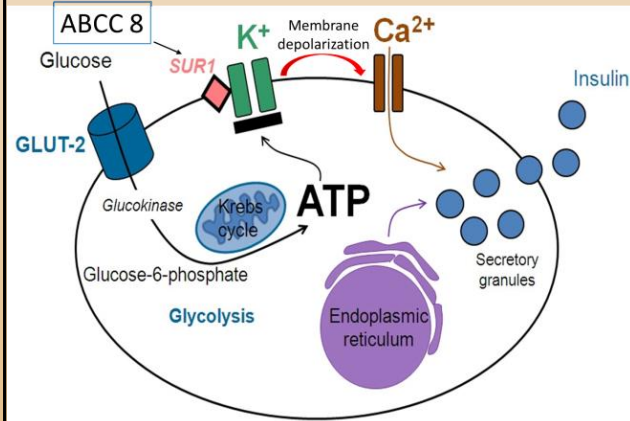


Rebound Hyperkalemia Post Subtotal Pancreatectomy in a Patient with Diffuse Congenital Hyperinsulinism from ABCC 8 Mutation

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Introduction

- Congenital hyperinsulinism (CHI) is a disease characterized by persistent insulin secretion unresponsive to hypoglycemia.
- One of the most common causes of CHI is mutation in the pancreatic ATP-sensitive potassium channel protein sulfonylurea receptor 1 (SUR1), encoded by ABCC 8 gene.



Case Description

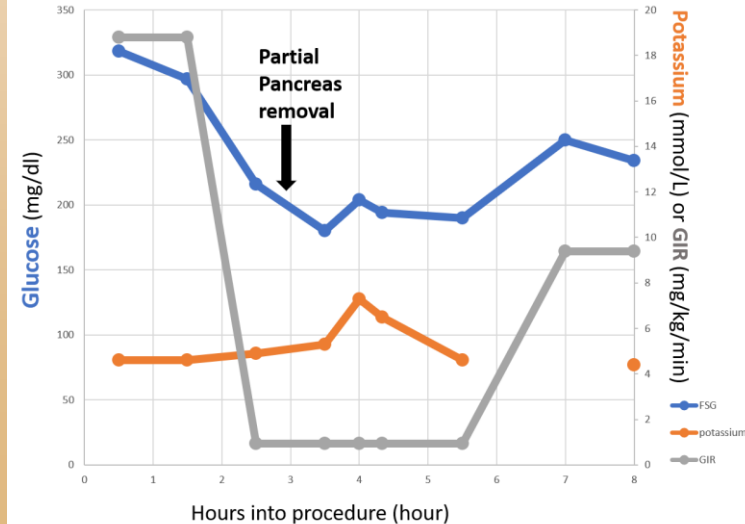
- Male born via uncomplicated vaginal delivery at 37-week gestation, birth weight 4.55kg; APGAR scores 8/9
- Family history: 2 siblings and 2 cousins with ABCC 8 homozygosity
- Initial glucose was 26; 10 when confirmed in the lab, requiring IV glucose infusion
- ABCC 8 gene sequencing confirmed ABCC 8 homozygosity

Operative Course

- At 15 days of life, patient presented for **subtotal pancreatectomy**
- General anesthesia with caudal catheter
- Arterial line for close hemodynamic and laboratory monitoring
- Within an hour after 97% of the pancreas was removed, we observed rebound hyperkalemia to 7.3 from baseline 4.6 without EKG changes

Postoperative

- K⁺ normalized without intervention while the patient's glucose infusion requirement increased
- Post-operative day 18, insulin/glucose balance was finally achieved
- POD 20 - discharged home on continuous tube feeds and octreotide



Discussion

- Due to the diffuse nature of the channelopathy, the disease is often unresponsive to medical therapy (diazoxide/octreotide)¹; thus pancreatic resection is a necessity.
- We postulate the patient's hyperkalemia after pancreatectomy was due to abrupt withdrawal of excessive endogenous insulin causing K⁺ efflux into the EC compartment.
- Post-operative potassium normalization is likely due to inflammation of the unresected pancreas causing excess insulin production discernible in the increased glucose demand postoperatively.
- Hyperkalemia-induced arrhythmia remains a potential complication as described by Austin, et al².
- We recommend potassium level and EKG monitoring while maintaining normoglycemia when caring for CHI patients perioperatively.

Conclusion

Healthcare providers need to be vigilant regarding potassium homeostasis and potential arrhythmia during pancreatectomy as well as maintain normoglycemia in spite of changing glucose level.

References

1. Arnoux JB, et al. Orphanet J Rare Dis 2011;6(63).
2. Austin JD, et al. Pediatr Crit Care Med 2008; 9(3):e17-9.
3. Burke MA, et al. Circ Res 2008; 102(2):164-76.