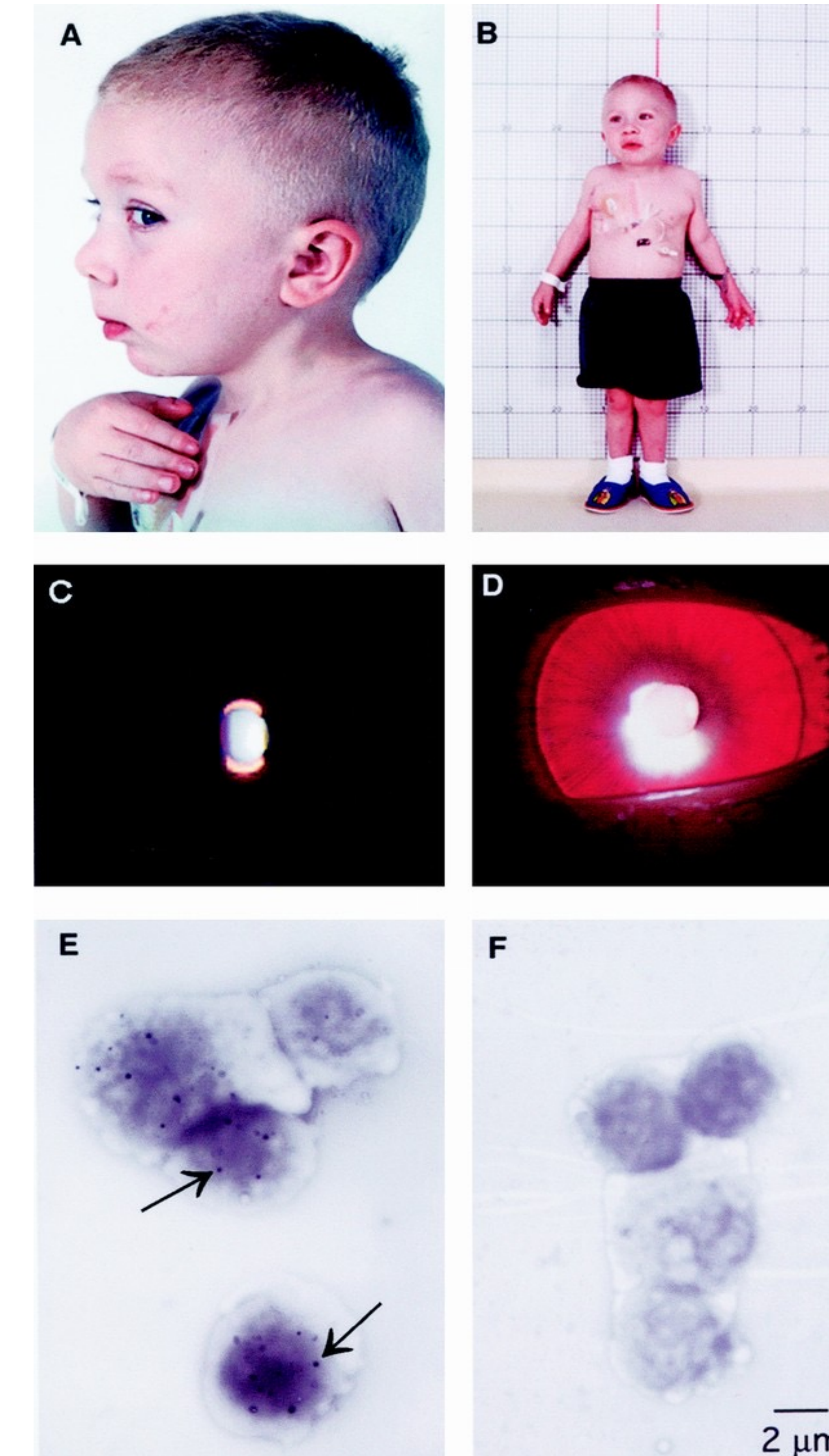


DIVERGENT ANALGESIA OUTCOMES AFTER SHARED DECISION-MAKING IN A PATIENT WITH COMPLEX BLEEDING DIATHESIS

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BACKGROUND

- Hermansky-Pudlak syndrome (HPS) is a rare autosomal recessive disease affecting lysosome-related organelles like platelet-dense granules and melanocytes
- To our knowledge, there have been case reports of 29 pregnancies in 15 women with HPS, most of whom received non-neuraxial labor analgesia¹
- There was only one patient who received neuraxial anesthesia when diagnosis of HPS was not known until after delivery¹
- Here we present a parturient with known HPS presenting for two separate L&D encounters
- We would like to highlight the role of shared decision-making in her labor analgesia plan



Huizing, M. *et al.* Nonsense Mutations in *ADTB3A* Cause Complete Deficiency of the β 3A Subunit of Adaptor Complex-3 and Severe Hermansky-Pudlak Syndrome Type 2. *Pediatr Res* **51**, 150–158 (2002).

PAST MEDICAL HISTORY

H/o anaphylaxis to platelets, Crohn's disease on chronic immunosuppression, chronic hypertension, migraine headaches, h/o provoked DVT in 2020 s/p apixaban

CLINICAL ENCOUNTER #1

- G1P0 at 36w5d presenting for IOL in the setting of IUGR
- Patient desired an epidural for labor analgesia
- Labs – Platelet 365K, PT 11.8s, aPTT 27s, INR 1.1
- Per hematology recommendation, she received 2U platelets after pre-medication, and ROTEM was done to check platelet function
- Epidural was placed successfully
- Patient progressed to have a successful vaginal delivery, EBL 250cc
- No complications from epidural placement or removal

CLINICAL ENCOUNTER #2

- G2P1001 at 33w2d presenting for an anesthesia consult and delivery planning
- Multidisciplinary discussion involving MFM, Hematology and OB Anesthesia
- Hematology recommendation – administer recombinant factor seven 90mcg/kg at time of delivery
- We had an in-depth discussion regarding risks and benefits of epidural placement
- Despite success with labor epidural during her prior pregnancy, patient opted for a fentanyl PCA
- She presented at 37w1d for IOL and proceeded to have a successful vaginal delivery
- She received 1g tranexamic acid during delivery, EBL 400cc
- Deferred postpartum tubal ligation due to inferior quality of analgesia

DISCUSSION

- This case series highlights the importance of shared decision-making while discussing options for labor analgesia
- Each L&D encounter is unique and it is important not to assume that the patient will accept the bleeding risk with an epidural despite having a successful experience in the past

REFERENCES

1. Obeng-Tuudah, D. et al. (2021), The presentation and outcomes of Hermansky-Pudlak syndrome in obstetrics and gynecological settings: A systematic review. Int J Gynecol Obstet, 154: 412-426.