



Touching lives with caring hands

# Intracranial haemorrhage in infant siblings: hereditary or acquired?

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17<sup>th</sup> INTERNATIONAL CHILD  
NEUROLOGY CONGRESS  
ANTALYA, TURKEY | OCTOBER 3-7, 2022

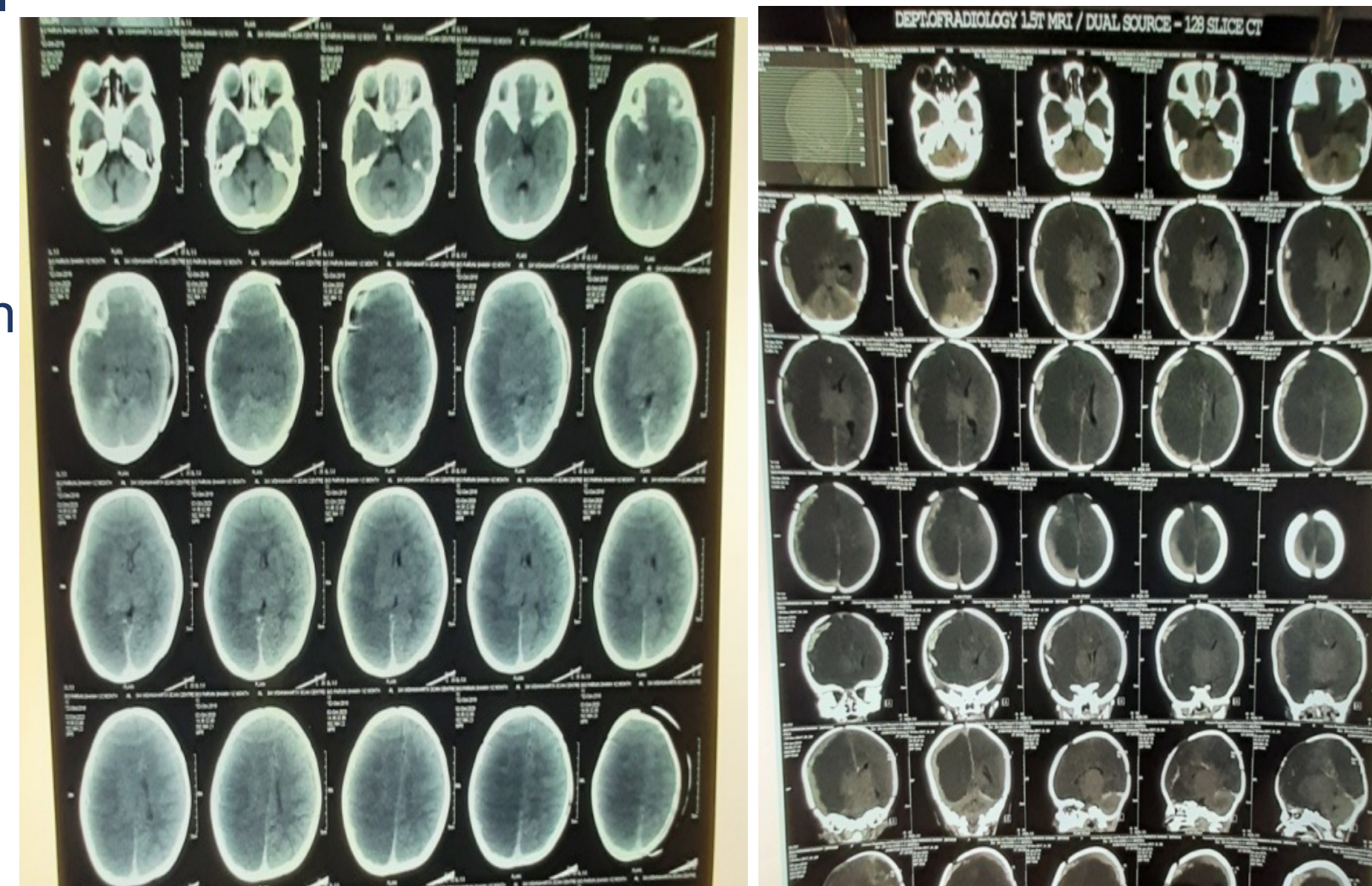
## INTRODUCTION

- ✓ Non traumatic intracranial hemorrhage (ICHs) is life threatening & uncommon in children.
- ✓ ICH in infantile age group needs to be managed with supportive therapy and a trial of Vit K injection for a reversible entity of Vitamin K Deficiency Bleeding (VKDB).
- ✓ The late onset VKDB present < 6th month of life.
- ✓ Incidence of 1/15000–1/20000 births
- ✓ We report two siblings born to a consanguineous couple with spontaneous ICH
- ✓ Despite consanguinity and sibling death from similar illness VKDB was considered

## MATERIALS & METHODS

- ✓ A 2-month-old female infant
  - ✓ vomiting episodes
  - ✓ left focal seizure
  - ✓ paucity of left side movements.
- ✓ On 3<sup>rd</sup> day of illness- worsening encephalopathy.
- ✓ At arrival :encephalopathic with poor cry, left hemiparesis and increased tone in left side.
- ✓ Born at village health facility with limited health care
- ✓ Term,vaginal delivery ,Birth Wt of 2.25kg.
- ✓ There was no h/o bleeding diathesis from umbilicus, injection site.
- ✓ Inv :Haemoglobin of 4.6 mg%, liver, renal function and coagulation profile(sent after FFP/Vit K administration ) were normal.
- ✓ A previous male baby presented elsewhere at 3 months succumbed to the bleed on d5 of illness. There were no documents avl with caregiver.

## NEUROIMAGING



➤ Index Case NCCT Head : bilateral frontoparietal SDH (3-4mm) without mass effect or midline shift.

➤ Deceased sibling NCCT Head : Rt sided hematoma over rt frontotemporal convexity, midline shift to lt side,

- ✓ Infant was managed with:

- ✓ Vit K Injection, component support,
- ✓ oxygen by head box ,IV antibiotics
- ✓ Antiseizure medications (ASM).

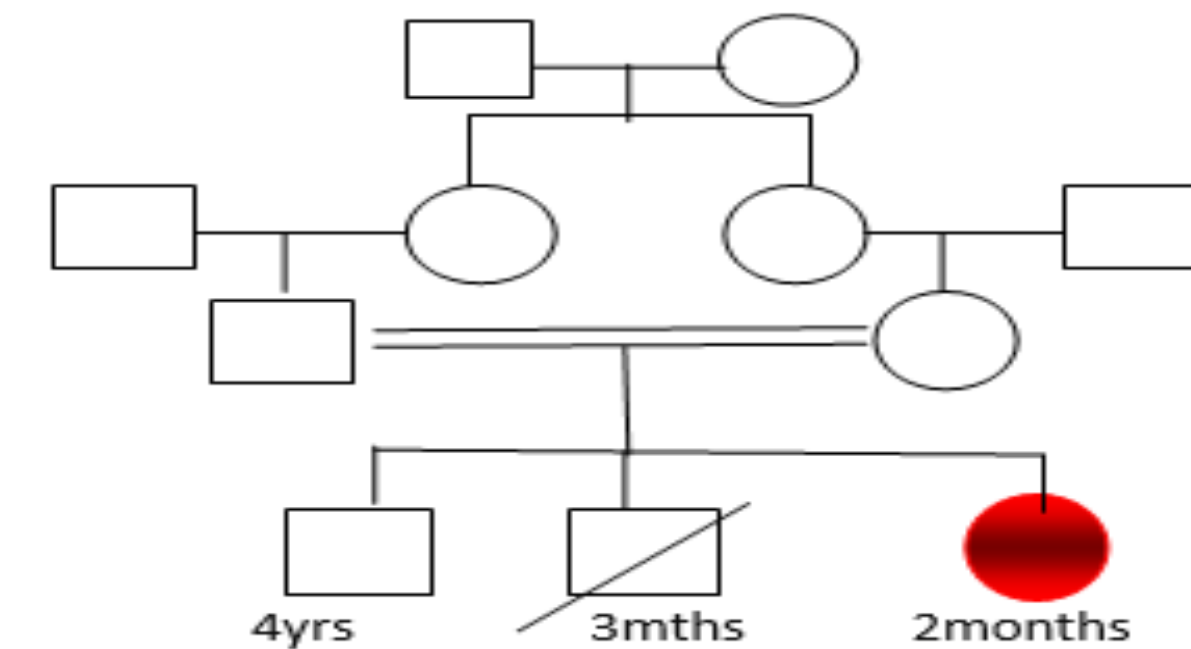
- ✓ Genetic testing keeping a possibility of a rare inherited haematological disorder owing to consanguinity, previous similar fatal case in sibling.

## RESULTS

- ✓ Protein Induced by Vit K Absence II (DCP PIVKA II) : A biomarker of VKDB = 1012.94 mAU/ml (17.36 -50.90). **20 times elevated**
- ✓ Factor IX activity-10.7%( 50-150%)
- ✓ Whole Exome sequencing(NGS): Normal

## FINAL DIAGNOSIS

1. Late Onset Vitamin K Deficiency Bleeding (VKDB)
2. Intra Cranial Haemorrhage  
Bilateral Sub Dural Hematoma (Right > Left)



## FOLLOW UP

- ✓ Child had a focal seizure on withdrawal of ASM at 6 months of age
- ✓ At 18 months gaining milestones with residual left hemiparesis.
- ✓ No bleeding recurrence
- ✓ Remains seizure free on single ASM
- ✓ EEG shows asymmetric Background activity and frequent Rt frontotemporal IEDs.

## DISCUSSION

- ✓ Most common causes of ICH are Vascular diseases (arteriovenous malformation, cavernoma, aneurysm, vasculitis, drugs) and blood disorder (ITP, VKDB, hemophilia, Hepatic disease ,DIC) .
- ✓ Clinical presentation and outcome may vary according to location, cause, rate of bleeding.
- ✓ Vitamin K is a cofactor for the activation of coagulation factors II, VII, IX, & protein C and S.
- ✓ VKDB is a disorder of hemostasis in which coagulation parameters are quickly corrected by vitamin K supplementation
- ✓ Three types of VKDB: early, classic and late.
- ✓ Exclusive breast feeding and cholestasis are closely associated with late onset VKDB
- ✓ Most reported cases of VKDB are ICH (82.7%)

- ✓ First siblings of VKDB with ICH to our knowledge.
- ✓ The siblings described here both had ICH:
  - ✓ First male baby presented elsewhere at 3 months and succumbed to the bleed.
  - ✓ The second female sibling index case : also at 2 months with ICH.

- ✓ PIVKAS are undercarboxyated precursor proteins of Vit K dependent Coag factors induced by Vit K deficiency
- ✓ More studies on subclinical VKDB needed to formulate guidelines for prevention so as to avoid life threatening bleeds.

## CONCLUSIONS

- ✓ Non traumatic ICH in infancy should raise suspicion of VKDB
- ✓ Must be empirically treated with IV Vitamin K awaiting investigations and blood component support as a life saving therapy.
- ✓ The index case is unique in terms of recurrence of VKDB in subsequently born siblings in a family
- ✓ Hence its prudent to administer Vitamin K to all newborns <6 mths of age as prophylaxis by focussed history if missed erroneously at birth.

## REFERENCES

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