# INTRODUCTION

- Neonatal McCune Albright syndrome (MAS) is very rare; presentation is more severe and widespread with multiple organ involvement.
- Hyperglycemia is very rare at presentation.
- Outcome of neonatal MAS is usually unfavorable with increased mortality.

## CASE PRESENTATION

- 11-day-old girl was admitted for weight loss and poor well-being.
- History of:
  - Respiratory distress
  - Elevated liver enzymes
  - ALT: 2376 U/L; AST: 875 U/L
- Neonatal sepsis
- Intrahepatic antibiotic regimen on first day of life

## PHYSICAL EXAMINATION

- Weight: 2315 g (-2.66 SDS)
- Head circumference: 32.6 cm (-1.88 SDS)
- BP: 70/30 mmHg
- Round facies, elongated philtrum, retromicrognatia
- Hyperpigmented macules
- Hypertrichosis

## LABORATORY ON ADMISSION

### 14th day
- Breast-fed neonate
- Persistent hyperglycemia (210 mg/dl)
- Insulin: 18.10 μIU/ml
- 0.5 units of subcutaneous NPH insulin (3x)

### 21st day
- Hypoglycemia
- Liver enzymes and renal function tests were elevated.

### 30th day
- Hypertrichosis
- Café au lait macules
- Hypocupremia and hypokalemia regressed
- Liver enzymes and renal function tests were elevated.

## CONCLUSION

- Neonatal MAS has a unique phenotype.
- Severity of initial hypercortisolism is a negative prognostic factor and early adrenalectomy is usually indicated.
- Three quarters adrenalectomy is an effective treatment for CS due to MAS.

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Presented at:
59ESPE
39th Annual Meeting Online
23-26 September 2021