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# Identification of critical illness-related corticosteroid insufficiency after congenital heart surgery with next generation sequencing

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## Background

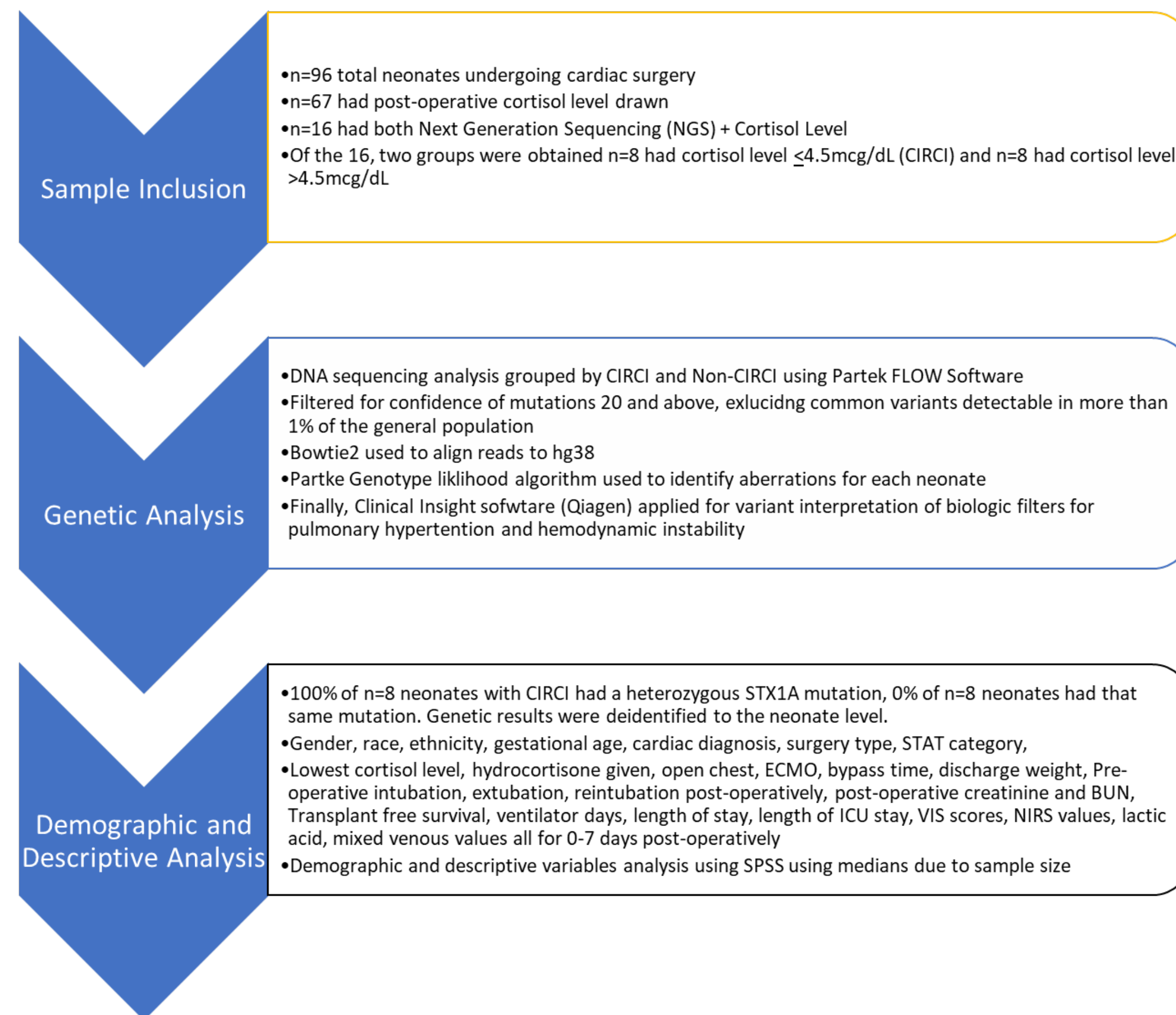
- Critical illness-related corticosteroid insufficiency (CIRCI) is common after cardiac surgery and is a cause of hemodynamic instability.

## Objectives

- Identify genetic abnormalities related to CIRCI in pediatric patients after congenital heart surgery.
- Describe differences in clinical outcomes between CIRCI and non-CIRCI groups.

## Methods

- Single-center retrospective study of neonates who underwent cardiac surgery between August 2018 to July 2020.
- Patients who had obtained postoperative cortisol levels and next gen DNA-sequencing (NGS).



## Results

- Seven gene mutations were present in 75-100% of patients with CIRCI.
- The CIRCI group had 100% incidence of heterozygous gene mutation on STX1A with splicing and loss of function compared to 0% incidence in the non-CIRCI group.

Table 1. Patient demographics and postoperative outcomes

	CIRCI	Non-CIRCI	P-value
<b>Patients (n)</b>	8	8	
<b>Age at surgery (days)</b>	10 (9 - 27)	9.5 (6 - 20)	0.62
<b>Gestational age (weeks)</b>	39 (38 - 41)	39 (38 - 40)	0.65
<b>Weight at surgery (kg)</b>	3.25 (2.73 - 4.8)	3.41 (2.6 - 4)	0.69
<b>CPB time (minutes)</b>	118.5 (81 - 177)	127 (107 -181)	0.80
<b>STAT Category</b>			
1	0	1	
2	0	0	
3	0	0	
4	4	4	
5	4	3	
<b>Cortisol (mcg/dL)</b>	2.15 (0 - 4.5)	5.95 (4.6 - 17.8)	<0.001
<b>CICU Length of stay (days)</b>	33 (5 - 125)	19.5 (8 - 45)	0.02
<b>Total length of stay (days)</b>	59.5 (49 - 307)	53.5 (21 - 343)	0.28
<b>Minimum SvO2 POD 0 (%)</b>	44.25 (14.7 - 77.4)	60.75 (23.7 - 80.3)	0.49
<b>Maximum VIS POD 0</b>	13 (0 - 22.5)	8 (5 - 45)	0.12
<b>Total mechanical ventilation (days)</b>	13 (3 - 22)	9.5 (3 - 18)	0.30

Data is represented in median (range).

CPB, cardiopulmonary bypass; CICU, cardiac intensive care unit; STAT, The Society of Thoracic Surgeons-European Association for Cardio-Thoracic Surgery score, SvO2, mixed venous oxygen saturation; POD, postoperative day; VIS, vasoconstrictor score.

## Conclusion

- Rapid testing for gene mutations has the potential to detect patients at risk for critical illness-related corticosteroid insufficiency with hemodynamic instability.
- Further research in larger patient cohorts is required to determine the statistical and clinical significance of these genetic abnormalities.