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Adrenal Crises in Children with Adrenal Insufficiency: Prevalence and Risk Factors

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Background

- Adrenal crises pose life-threatening emergencies that may occur in patients with AI, either at presentation due to delay in diagnosis or under appropriate steroid replacement therapy due to coexisting illness or non-compliance
- Studies on the rate of AC in children with AI are scarce, and they were done mainly on children with CAH

Objectives

To assess the prevalence of AC and mortality rate in children with AI and to evaluate the risk factors for their development

Methods

- The study included children diagnosed with AI between 1990 and 2017 at 4 pediatric endocrinology units in Israel
- Data were retrieved retrospectively from the patients' files, and they included demographic factors, clinical information (age at diagnosis, specific diagnosis, and clinical presentation during AC), and details about therapy regimens (type, dosage, and parental guidance)
- AC was defined as a clinical deterioration precipitated by a glucocorticoid-deficient state, or clinical improvement after administration of glucocorticoids with at least 2 of the following: severe fatigue, nausea and vomiting, somnolence, hypotension, hyponatremia, hyperkalemia, or hypoglycemia

Results

- 120 children (73 boys, 47 girls)
- 904 patient years (95% CI, 792-1018)
- Median age at diagnosis was 0.25 years (0-17.5)
- Median age at study was 11.5 years (0.3-25)
- The frequency of AC was 3.4 per 100 patient years
- 52% of AC events occurred at presentation, 42% followed infection disease, and 6% were the result of noncompliance with treatment
- Preventive measures of carrying emergency card or a possession of an emergency kit did not arrest the development of AC
- There was no mortality

Risk factors for developing AC

Risk factor	OR	95%CI
Age younger than 5 years at diagnosis	5.3	1.2-25
Primary AI	4.5	1.3-16.7
Specific diagnosis: SW CAH, AHC, Addison's disease, ALD		
Mineralocorticoid treatment	19.3	4.3-175
Recurrent hospital admissions		

The Authors have nothing to disclose

Children who developed AC vs. those who did not

	Children that developed AC (n = 26)	Children that did not develop AC (n =94)	*P*
Age at diagnosis (yr)	0.96 ± 2.35	3.92 ± 5.13	0.04
Gender			0.069
Male, n	20 (27.4%)	53 (72.6%)	
Female, n	6 (12.2%)	41 (87.8%)	
Gender (X-linked cases excluded)			0.31
Male, n	13 (20.3%)	51 (79.7%)	
Female, n	6 (12.2%)	43 (87.8%)	
AI diagnosis			0.016
Primary, n	23 (28%)	59 (72%)	
Secondary, n	3 (7.9%)	35 (92.1%)	
Specific AI diagnosis			<0.001
AHC, ALD and Addison, n	9 (64.3%)	5 (35.7%)	
Salt wasting CAH, n	12 (37.5%)	20 (62.5%)	
Simple virilizing CAH, n	2 (5.6%)	34 (94.4%)	
Secondary (idiopathic), n	3 (14.3%)	18 (85.7%)	
Secondary (tumor), n	0 (0%)	17 (100%)	
Hormones values at diagnosis (nmol/L)			
Basal cortisol, mean ± SD	128.2 ± 121.6	183.6 ± 166.2	0.13
Peak cortisol, mean ± SD	271.0 ± 256.0	289.9 ± 207.8	0.75
MC therapy needed, n	24 (92.3%)	2 (3.3%)	<0.001
MC therapy dosage (mg)	0.1 ± 0.05	0.09 ± 0.04	0.46
HC therapy dosage (mg/m^2/day)	13.3 ± 4.8	11.9 ± 5.3	0.2
Emergency card, n	25 (96%)	79 (84%)	0.18
Emergency kit, n	13 (50%)	43 (45.7%)	0.82
Hospital admissions, mean ± SD	2.34 ± 1.41	1.3 ± 1.35	<0.001

Summary

- Diagnosis and long-term management of patients with adrenal insufficiency remains a challenge
- Since significant percent of AC events occurred at presentation measures to increase the awareness to signs and symptoms of AI among primary care physicians are paramount importance
- Efforts to prevent AC should be focused in younger patients especially those with primary AI