

ESPE Code	<b>Diagnosis of disorders of the adrenal gland according to the European society of Pediatric Endocrinology (ESPE)</b>	OMIM	ICD-10
<b>8A</b>	<b>PRIMARY ADRENAL INSUFFICIENCY</b>		
<b>8A.1</b>	<b>Congenital adrenal hyperplasia (CAH)</b>		E25
8A.1a	Cholesterol side-chain cleavage deficiency (lipoid CAH) (P450sc)	#201710	E25.0
8A.1b	3beta-Hydroxysteroid dehydrogenase deficiency	2214158 4150	E25.0
8A.1c	21-Hydroxylase deficiency (P450c21)	201910	E25.0
8A.1c.1	Salt-wasting 21-OHD		
8A.1c.2	Simple virilizing 21-OHD		
8A.1c.3	Non-classical 21-OHD		
8A.1d	11beta-Hydroxylase deficiency <sup>7</sup>	#202010	E25.0
8A.1e	P450c11AS deficiency	*124080	E25.0
8A.1e.1	Corticosterone methyl oxidase deficiency type I		E25.0
8A.1e.2	Corticosterone methyl oxidase deficiency type II		E25.0
8A.1e.3	Glucocorticoid suppressible hypertension	#103900	E25.0
8A.1f	17alpha-Hydroxylase/17/20 lyase deficiency	#202110	E25.0
8A.1g	P450 oxidoreductase deficiency (may be part of Antley Bixler congenital malformation syndrome)	#201750	E25.0
8A.1h	Glucocorticoid receptor defect	138040	E25.0
8A.1z	Other disorders, unspecified		E25.9
<b>8A.2</b>	<b>Other causes of adrenal insufficiency</b>		
8A.2a	Congenital adrenal hypoplasia	#300200	Q89.1
8A.2a.1	DAX-1 (NROB1) mutation <sup>14</sup> (also classified as 6A.3a.2 if combined with hypogonadotrophic hypogonadism)		
8A.2a.2	Idiopathic		
8A.2b	Adrenoleukodystrophy (Schilder's disease) and adrenomyeloneuropathy	#202370	E71.3
8A.2c	Primary xanthomatosis (Wolman's disease)	278000	Q89.1
8A.2d	Familial glucocorticoid deficiency (ACTH insensitivity, hereditary unresponsiveness to ACTH)	#202200	Q89.1
8A.2e	Triple A (Allgrove) syndrome	#231550	Q89.1
8A.2f	Pseudohypoaldosteronism, type 1	#264350	Q89.1
8A.2z	Other syndromes, unspecified		Q89.1
<b>8A.3</b>	<b>Autoimmune adrenalitis (Addison's disease)</b>		E27.1
8A.3a	Disorders classified elsewhere: <i>Part of autoimmune polyglandular syndromes: – Type 1 classified as 14C.4a – Type 2 classified as 14C.4b Other types (see 14C.4c, d and e) Part of Steinert syndrome (14B.35)</i>		
8A.3b	Isolated		
<b>8A.4</b>	<b>Infections</b>		E27.8
8A.4a	Tuberculosis		A18.7, E35.1
8A.4b	Fungal infections		E27.4
8A.4c	Bacterial sepsis		E27.4
8A.4d	AIDS		E27.4
8A.4y	Other specified infections		

<b>8A.5</b>	<b>Haemorrhage</b>		
8A.5a	Associated with meningococcal infection (Waterhouse-Friedrichsen syndrome)		A39.1, E35.1
8A.5y	Other specified causes		E27.4
8A.5z	Idiopathic		E27.4
<b>8A.9</b>	<b>Idiopathic</b>		E27.4
<b>8B</b>	<b>SECONDARY ADRENAL INSUFFICIENCY</b>	Classified elsewhere: 6A.1	
<b>8C</b>	<b>ADRENAL EXCESS</b>		
<b>8C.1</b>	Glucocorticoid excess (Cushing syndrome) [primary] [secondary 14C.1]		E24
8C.1a	Caused by ACTH excess		E24.0
8C.1a.1	Cushing's disease (ACTH-producing pituitary adenoma)	219090	E24.0
8C.1a.2	CRF excess		E24.3
8C.1a.3	Ectopic ACTH syndrome		E24.3
8C.1b	Increased peripheral glucocorticoid production		E24.8
8C.1b.1	ACTH-independent macronodular adrenal hyperplasia	#219080	E24.8
8C.1b.2	Multinodular adrenal hyperplasia, isolated or as part of the Carney complex <sup>31</sup>	#160980	E24.8
8C.1b.3	Adrenal adenoma		E24.8
8C.1b.4	Adrenal carcinoma		E24.8
8C.1c	Glucocorticoids from other sources		E24.8
8C.1c.1	Iatrogenic Cushing syndrome		E24.2
8C.1c.z	Other		E24.2
<b>8C.2</b>	<b>Virilising and feminising adrenal tumours</b>		
<b>8C.3</b>	<b>Mineralocorticoid excess</b>		E26.0
8C.3a	Conn syndrome		E26.0
8C.3b	Substances with mineralocorticoid action (liquorice)		E26.0
<b>8D</b>	<b>DISORDERS OF THE ADRENAL MEDULLA</b>		
<b>8D.1</b>	<b>Adrenal medulla defect</b>		
8D.1a	Haemorrhage		A39.1, E35.1
8D.1b	Infection		E35.1
8D.1c	Tumour		E35.1
8D.1z	Idiopathic		E35.1
<b>8D.2</b>	<b>Primary tumour in adrenal medulla</b>		
8D.2°	Phaeochromocytoma		M8700/0
8D.2b	Neuroblastoma		M9500/3