ESPE Code	***Diagnosis of pituitary gland and hypothalamus according to the European society of Pediatric Endocrinology (ESPE)***	**OMIM**	**ICD-10**
6A	**DEFICIENCIES OF ANTERIOR PITUITARY HORMONES**		
6A.0	**Disorders classified elsewhere**		
6A.1	**ACTH deficiency**		E23.0
6A.1a	Congenital isolated ACTH deficiency	#201400	
6A.1b	Congenital ACTH deficiency in combination with other pituitary deficiencies		
6A.1c	Acquired hypothalamic-pituitary ACTH deficiency, e.g. by longterm glucocorticoid therapy		
6A.2	**TSH deficiency**		E03.9
6A.2a	Congenital isolated TSH deficiency		E23.0
6A.2a.1	– Known genetic defect (TSH-beta, TRHR, TRH, other)	#275100 +188545 +275120	E23.0
6A.2a.2 –	Unknown origin		E23.0
6A.2b	Congenital TSH deficiency in combination with other pituitary deficiencies		E23.0
6A.2b.1	– Secondary (pituitary) hypothyroidism		E23.0
6A.2b.2	– Tertiary (hypothalamic) hypothyroidism		E23.0
6A.2c	Acquired hypothalamic-pituitary hypothyroidism		E23.0
6A.3	**Gonadotrophin deficiency (hypogonadotrophic hypogonadism)**		E23.0
6A.3°	X-linked inheritance		E23.0
6A.3a.1	– Isolated hypogonadotrophic hypogonadism and anosmia (X-linked form of Kallmann syndrome) (KAL1 mutation); [primary] [secondary 14B.16]	308700	E23.0
6A.3a.2	– Isolated hypogonadotrophic hypogonadism and adrenal hypoplasia congenita (DAX1 mutation) (*also classified as* 8A.2a.1)		E23.0
6A.3b	Autosomal inheritance: autosomal forms of Kallmann syndrome		E23.0
6A.3b.1	– Inactivating mutation of FGFR1 (KAL2)	#147950	
6A.3b.2	– Inactivating mutation of PROKR2 (KAL3)	244200	
6A.3b.3	– Inactivating mutation of PROK2 (KAL4)	610628	
6A.3b.4	– Inactivating mutation of GnRHR	*138850	E23.0
6A.3b.5	– GPR54 mutation	*604161	E23.0
6A.3b.6	– KISS mutation	*603286	E23.0
6A.3b.7	– Prohormone convertase 1 mutation	600955	E23.0
6A.3b.8	– Leptin 1 or leptin receptor mutation	*164160 *601007	E23.0
6A.3b.9	– LHR mutation (fertile eunuch syndrome)	#228300	
6A.3b.10	– Isolated LH deficiency	152780	E23.0
6A.3b.11	– Isolated FSH deficiency	#229070	E23.0
6A.3b.88	– Other specified gene mutations, including NELF, etc.	*608137	E23.0
6A.3c	Hypogonadotrophic hypogonadism in combination with other pituitary deficiencies		E23.0
6A.3c.1	– Known genetic defect, e.g. PROP1, LHX3, HESX1	*601538 *600577 *601802	E23.0
6A.3c.2	– Unknown genetic defect		E23.0

6A.3d	Hypogonadotrophic hypogonadism in combination with dysmorphic syndromes, e.g. Prader-Willi-Labhart syndrome [primary 14B.25], Rieger syndrome [primary 14B.28]		E23.0
6A.3z	Hypogonadotrophic hypogonadism, isolated, unspecified		E23.0
6A.4	**Prolactin deficiency**		
6A.4a	Isolated prolactin deficiency	264110	E23.0
6A.4b	Prolactin deficiency in combination with other pituitary deficiencies		E23.0
6A.4b.1	POU1F1 mutation	173110	E23.0
6A.4b.2	PROP1 mutation	*601538	E23.0
6A.4b.8	Other specified gene mutations		E23.0
6A.4b.9	Unknown origin		E23.0
6B	**OVERPRODUCTION OF ANTERIOR PITUITARY HORMONES**		
6B.0	**Disorders classified elsewhere** *ACTH-producing adenoma (Cushing's disease)* (8C.1) *Growth hormone-producing adenoma (synonyms: acromegaly, pituitary gigantism)* (2B.1)		
6B.1	**TSH-producing adenoma**		E05.9
6B.2	**Gonadotrophin-producing adenoma**		
6B.3	**Prolactin overproduction**		
6A.3a	Prolactinoma		
6A.3b	Hyperprolactinaemia of other cause (e.g. pituitary stalk lesion, primary hypothyroidism)		E22.1
6C	**CENTRAL D IABETES INSIPIDUS** [primary 13A.1]		
6D	**HYPOTHALAMIC DYSFUNCTION, NOT CLASSIFIED E LSEWHERE**		E23.3
6D.0	**Disorders classified elsewhere** *Obesity* (5D)		
6D.8	**Other specified functional changes**		
6D.9	**Other functional changes, unspecified**		
SECTION 2 AETIOLOGICAL CLASSIFICATION			
6E	**CONGENITAL DISORDERS**		
6E.1	**Congenital CNS malformations**		Q04
6E.1°	Septo-optic dysplasia [primary] [secondary 14B.30]	#182230	Q04.4
6E.1b	Other midline defects, e.g. cleft palate, central maxillary incisor syndrome, EEC syndrome (ectodactyly ectodermal dysplasiaclefting syndrome)		Q04.8
6E.1c	Ectopic neurohypophysis, absent infundibulum and hypoplastic adenohypophysis		Q04.8
6E.1d	Hamartoma	241800	Q85.9
6E.1z	Other congenital CNS malformations, unspecified		Q04.9

6E.2	**Congenital hypothalamic-pituitary disorders associated with syndromes**		
6E.2a	Disorders classified elsewhere: *Prader-Willi(-Labhart) syndrome* (14B.25) *Rieger syndrome* (14B.28)		
6E.2b	Chromosomal disorders		
6E.2c	Chromosomal instability syndromes		
6E.2d	Empty sella syndrome		
6E.2y	Other specified syndromes		
6E.2z	Other syndromes, unspecified		
6F	**ACQUIRED DISORDERS**		
6F.1	**Neoplasms**		C71.9
6F.1a	Tumours of the pituitary/hypothalamic region		
6F.1a.1	Craniopharyngioma		
6F.1a.2	Nonfunctional pituitary adenomas		
6F.1a.3	Other benign structures		
6F.1a.4	Isolated glioma		
6F.1a.5	Glioma as part of neurofibromatosis I (von Recklinghausen's disease, 14B.27)		
6F.1a.6	Germinoma, dysgerminoma		
6F.1a.7	Leukaemia, lymphoma		
6F.1a.8	Other neoplasms, specified		
6F.1b	Tumours outside the pituitary/hypothalamic region		
6F.1b.1	Pinealoma		
6F.1b.2	Isolated glioma		
6F.1b.3	Glioma as part of neurofibromatosis I (von Recklinghausen's disease, 14B.27)		
6F.1b.4	Germinoma, dysgerminoma		
6F.1b.5	Medulloblastoma		
6F.1b.6	Leukaemia, lymphoma		
6F.1b.8	Other specified neoplasms		
6F.2	**Inflammatory/infiltrative**		
6F.2a	Langerhans cell histiocytosis	604856	D76.0 D76.3
6F.2b	Systemic lupus erythematodes	#601744 #152700	
6F.2c	Neurosarcoidosis	#181000	
6F.2d	Lymphocytic neurohypophysitis		
6F.2e	Haemochromatosis	#602390	
6F.3	**Infectious**		
6F.3a	Meningitis		G00
6F.3b	Encephalitis		G04, G05
6F.3c	Abscess of pituitary		G06.0
6F.3d	Congenital infection		G09
6F.4	**Traumatic injury**		
6F.4a	CNS surgery		G97
6F.4b	Head trauma		S06
6F.4c	Hypoxic injury		G97.8
6F.5	**Iatrogenic**		
6F.5a	Irradiation		T66
6F.5b	Drugs, e.g. chemotherapy		E23.1

6F.6	**Secondary to psychiatric disorders**		
6F.6a	Anorexia nervosa		F50.0
6F.6b	Emotional deprivation		
6F.6c	Other (specified)		
6F.7	**Idiopathic**		