内分泌分野

ICD-11 alpha-version (provisonal) by Inernal Medicine TAG Endo/Metab

Disorders of thyroid gland

Congenital hypothyroidism

- Congenital hypothyroidism due to iodine deficiency
  - Congenital iodine-deficiency syndrome, neurological type
  - Congenital iodine-deficiency syndrome, myxoedematous type
  - Congenital iodine-deficiency syndrome, mixed type
  - Congenital iodine-deficiency syndrome, unspecified
- Permanent congenital hypothyroidism with diffuise goitre
  - Dyshormogenetic hypothyroidism
  - Congenital hypothyroidism due to resistance to thyroid hormones
- Pendred's sundrome
- Permanent congenital hypothyroidism without goitre
  - Congenital thyroid dysgenesis
  - Primary congenital hypothyroidism due to resistance to TSH

Iodine-deficiency-related thyroid disorders and allied conditions

- Iodine-deficiency-related diffuse (endemic) goitre
- Iodine-deficiency-related multinodular (endemic) goitre
- Iodine-deficiency-related (endemic) goitre, unspecified
- Other iodine-deficiency-related thyroid disorders and allied conditions

Subclinical iodine-deficiency hypothyroidism

Acquired hypothyroidism

- Hypothyroidism due to medicaments and other exogenous substances
- Postinfectious hypothyroidism
- Autoimmune thyroiditis
- Other rare specified hypothyroidism
- Hypothyroidism, unspecified

Other nontoxic goitre

- Nontoxic diffuse goitre
- Nontoxic single thyroid nodule
- Nontoxic multinodular goitre
- Other specified nontoxic goitre
- Nontoxic goitre, unspecified

Hyperthyroidism [thyrotoxicosis]

- Hyperthyroidism (thyrotoxicosis) with diffuse goitre
- Hyperthyroidism (thyrotoxicosis) with toxic single thyroid nodule
- Hyperthyroidism (thyrotoxicosis) with toxic multinodular goitre
- Hyperthyroidism (thyrotoxicosis) from ectopic thyroid tissue
- Thyrotoxicosis factitia
- Thyroid crisis or storm
- Hyperthyroidism (thyrotoxicosis), familial
  - Hyperthyroidism, familial, due to mutations in TSH receptor
  - Selective pituitary resistance to thyroid hormone
- Other Hyperthyroidism (thyrotoxicosis)
  - TSH-secreting pituitary adenoma
  - Overproduction of throid-stumulating hormone
- Hyperthyroidism (thyrotoxicosis), unspecified

Thyroiditis

- Acute thyroiditis
- Subacute thyroiditis
- Chronic thyroiditis with transient thyrotoxicosis
- Autoimmune thyroiditis
- Drug-induced thyroiditis
- Other rare specified chronic thyroiditis
  - Riedel thyroiditis
- Thyroiditis, unspecified

Other disorders of thyroid

- Hypersecretion of calcitonin
  - C-cell hyperplasia of thyroid
  - Thyroid carcinoma, medullary
- Thyroid tumors
  - Throid carcinoma, papillary or follicular
  - Thyroid carcinoma, anaplastic
- Other specified disorders of thyroid
  - Haemorrhage of thyroid
  - Infarction of thyroid
  - Sick-euthyroid syndrome
  - Generalized resisitance to thyroid hormone
- Disorder of thyroid, unspecified

Disorders of other endocrine glands

Disorders ot the parathyroid glands and phosphocalcic metabolism

- Hypoparathyroidism
  - Idiopathic hypoparathyroidism
    - Autoimmune hypoparathyroidism
  - Pseudohypoparathyroidism
    - Pseudohypoparathyroidism, type 1A
    - Pseudohypoparathyroidism, type 1B
    - Pseudohypoparathyroidism, type 1C
    - Pseudohypoparathyroidism, type 2
    - Psudopseudohypoparathyroidism,
  - Hypoparathyroidism due to agenesis or dysgenesis of the parathyroid glands
  - Primary hypoparathyroidism
  - Secondary hypoparathyroidism
    - Hypothyroidism due to hypomagnesemia
    - Hypoparathyroidism due to destruction of the parathyroid glands
    - Hypoparathyroidism after iodine thyroid ablation
    - Hypoparathyroidism due to external radiation
    - Infiltrative disorder with hypoparathyroidism
  - Other rare hypoparathyroidism
    - Autosomal dominant hypocalcemia (ADH)
  - Hypoparathyroidism, unspecified

- Hyperparathyroidism and other disorders of parathyroid gland
  - Non-familial primary hypoparathyroidism
    - Hyperplasia of parathyroid
    - Parathyoid adenoma
    - Parathyroid carcinoma
  - Primary hyperparathyroidism neonatal severe
  - Primary hyperparathyroidism, familial
    - Parathyroid adenoma, familial
    - Multiglandular hyperplasia, familial
    - Hyperparathyroidism, familial, isolated (FIHPT)
    - Hyperparathyroidism-jaw tumor syndrome (HPT-JT)
  - Secondary hyperparathyroidism, not elesewhere classified
  - Other rare hyperparathyroidism
    - Hyperparathyroidism, unspecified
  - Other rare specified disorders of parathyroid gland
  - Disorder of parathyroid gland, unspecified

Disorders of the pituitary hormones system

- Hypersecretion of pituitary hormones
  - Acromegaly and pituitary gigantism
    - Somatotroph adenoma
  - Hyperprolactinaemia
    - Prolactinoma
    - Drug-induced
  - Syndrome of inappropriate secretion of antidiuretic hormone (SIADH)
    - Pituitary inappropriate secretion of antidiuretic hormone
    - Ectopic inappropriate secretion of antidiuretic hormone
  - Other hypersecretion of pituitary hormones
  - Hypersecretion of pituitary hormones, unspecified
- Hyposecretion of pituitary hormones and other disorders of hypothalamus and/or the pituitary hormones
  - Combined pituitary deficiency
  - Congenital Hypogpnadotropic hypogonadism
    - Hypogonadotropic hypogonadism normosmic
    - Hypogonadotropic hypogonadism amosmic
  - Nonacuired isolated growth hormone deficiency
  - Growth hormone deficiency with or without pituitary stalk interruption syndrome (PSIS)
    - Idiopathic growth hormone deficiency
    - Genetic growth hormone deficiency
  - Short stature due to defect in growth hormone receptor or post-receptor pathway
    - Short stature dut to growth hormone resistance
    - Growth delay due to insulin-like growth factor 1 deficiency
    - Growth delay due to insulin-like growth factor 1 resistance
  - Acquired (or secondary) hypopituitarism
    - Drug-induced hypopituitarism
    - Tumoral hypopituitarism
    - Infectious hypopituitarism
    - Autoimmune hypopituitarism
    - Vascular hypopituitarism
    - Sheehan's sundrome

- Metabolic (storage-deseases)
- Seconfdary to a granulomatous disease
- Traumatic hypopituitarism

Central diabetes insipidus

Hypothalamic dysfunction, not elsewhere classified

- Diencephalic syndrome

Other rare disorders of pituitary gland

- Abscess of pituitary
- Adipospgenital dystrophy
- Isolated ACTH deficiency (excluding congenital)
- Pituitary stalk interruption syndrome (PSIS) NOS
- Isolated FSH deficiency

Disorder of pituitary gland, unspecified

Disorders of the adrenal glands and adrenal hormonal system

Cushing's syndrome

ACTHdependent Cushing's syndrome

- Cushing's disease
- Cushing's syndrome secondary to ectopic ACTH secretion

ACTH-independent Cushing's syndrome

- Adrenal adenoma
- Cortical adrenal carcinoma

Nelson's syndrome

Drug-induced Cushing's syndrome

Psudo-Cushing's syndrome

- Alcohol-induced pseudo-Cushing's syndrome
- Pseudo-Cushing's syndrome of othert causes

Other rare Cushing's syndrome

Cushing's syndrome, unspecified

Adrenocortical Insufficiency

Chronic primary adrenocortical insugfficiency, non congenital

Congenital adrenocortical insufficiency

- Congenital isolated ACTH deficiency
- Adrenal hypoplasia, familial
- Adrenal hypoplasia, congenital, of matemal cause
- Adrenal hypoplasia, congenital, cytomegalic

Acute adrenocortical insufficiency

- Interruption of corticosteroid therapy
- Adrenal hemorrhage
- Adrenal infarction

Drug-induced chronic adrenocortical insufficiency

Other specified rare adrenocortical insufficiency

Admocortical insufficiency, unspecified

Adrenogenital disorders

Congenital adrenal hyperplasia

- Classical congenital adrenal hyperplasia (salt-losing)
- Non-classical congenital adrenal hyperplasia (non salt-losing)

Other adrenogenital disorders

- Premature adrenarche
- Drug-induced adrenogenital disorders
- Congenital 46, XX sex development disorders induced by (adrenal or ovarian) androgens or matemal origin

Adrenogenital disorder, unspecified

Hyperaldosteronism

Primary hyperaldosteronism, non familial

- Conn's adenoma
- Primary aldosteronism due to adrenal hyperplasia (bilateral)

Familal hyperaldosteronism type 1

Familal hyperaldosteronism type 2

Secondary hyperaldosteronism

Other hyperaldosteronism

Hyperaldosteronism, unspecified

Hypoaldosteronism

Hypoaldosteronism, familial

Other hypoaldosteronism

Hypoaldosteronism, unspecified

Other disorders od adrenal hormones

Hypersecretion of adrenomedullary hormones

- Pheochromocytoma
- Secreting paraganglioma
- Adrenomedullary hyperplasia

Other rare specified disorders of adrenal gland

Adrenal incidentaloma

Disorder of adrenal gland, unspecified

Disorders of the gonadal hormones system

Ovarian dysfunction and ovarian hormones disorders

Estrogen excess.

Polycystic ovarian syndrome

Other ovarian androgen excess

Drug-induced androgen excess

Tumoral androgen excess

Premature ovarian failure

Gonadal dysgenesis XX (FSH-resistant ovaries, Hypergonadotropic ovarian dysgenesis, resistant ovary syndrome)

Other rare ovarian dysfunction

Ovarian hyperstimulation syndrome

Ovarian dysfunction, unspecified

Testicular dysfunction and testosterone-related disorders

Testicular hyperfunction

Testicular hypofunction

Defective biosynthesis of testicular androgen (Testicular hypogonadism)

46, XY sex development disorder due to a defect in testosterone metabolism

5-Alpha-reductase deficiency

Leydig cell hypoplasia

Androgen resistance syndrome

Complete periheral androgen resistance syndrome

Partial peripheral androgen resistance syndrome (Reifenstein's syndrome)

Dysgenetic 46, XY sex development disorder

Other rare testicular dysfunction

Testicular dysfunction, unspecified

Disorders of puberty, not elsewhere classified

Delayed puberty

Peripheral precocious puberty

Testoxicosis

Central precocious puberty

Other disorders of puberty

Disorder of puberty, unspecified

Polyglandular dysfunction

Autoimmune polyglandular failure

Type 1 autoimmune polyendocrinopathy

Type 2 autoimmune polyendocrinopathy (Schmidt's syndrome)

Polyglandular hyperfunction

Carney complex

Carney-Stratakis syndrome

Carney trad

Multiple endocrine neoplasia, type 1

Multiple endocrine neoplasia, type 2

Other polyglandular dysfunction

Polyglandular dysfunction, unspecified

Other endocrine disorders

Carcinoid syndrome

Neuroendocrine tumors NOS

Ectopic hormone secretion, not elsewhere classified

Short stature, not elsewhere classified

Constitutional tall stature

Other rare specified endocrine disorders

Endocrine disorder, unspecified

To be classified under immunological diseases

Diseases of thymus

Persistent hyperplasia of thymus

Abscess of thymus

Other diseases of thymus

Disease of thymus, unspecified

Diabetes Mellitus

- .0 With coma
  - Diabetic coma
  - Hyperglycemic hyperosmolar coma
  - Hypoglycemic coma
- .1 With acidosis
  - Ketoacidosis
  - Lactic acidosis
  - Metabolic acidosis
- .2 With diabetic nephropathy
  - Stage 1 (very early stage)
  - Stage 2 (Microalbuminuria)
  - Stage 3 (Macroalbuminuria)
  - Stage 4 (end-stage renal disease)
  - Stage 5 (dialysis)
- .3 With diabetic retinopathy
  - Retinopathy
    - simple
    - preproliferative
    - proliferative
  - Macular edema
  - Cataract
  - Glaucoma
  - Blindness
- .4 With nerological complications
  - polyneuropathy
    - sensory
    - antonomiorthostatic hypotension
      - gastroparesis
    - motor
  - mononeuropathy
    - Cranial nerve dysfunction
    - Trunk/Extremity nerve dysfunction
    - amyotrophy
  - genitourinary
    - sexual dysfunction
    - uropathy
- .5 With macrovascular complications
  - Coronary artery disease
  - Peripheral arterial disease
  - Cerebrovascular disease
- .6 With other specified complications
  - Diabetic foot
    - arthropathy
    - gangrene
    - ulcer
    - lower extremity amputation
  - Diabeteic skin lesion
  - Periodontal disease
- .7 With multiple complications
- .8 With unspecified complications
- .9 Without complications

E10 Type 1 diabetes mellitus

- Immune-related
  - rapidly progressive form
  - slowly progressive form (SPIDDM, LADA)
  - fulminant type
- Idiopathic

E11 Type 2 diabetes mellitus

E13 Other specific types of diabetes mellitus

- Genetic defects of β cell function
  - Chromosome 12, Hepatocyte nuclear factor HNF-1α(MODY3)
  - Chromosome 7, Glucokinase (MODY2)
  - Chromosome 20, HNF-4α(MODY1)
  - Chromosome 13, Insulin promoter factor-1 (IPF-1;MODY4)
  - Chromosome 17, HNF-1β(MODY5)
  - Chromosome 2, NeuroD1 (MODY6)
  - Mitochondrial DNA
  - Neonatal diabetes
  - Others
- Genetic defects in insulin action
  - Type A insulin resistance
  - Leprechaunism
  - Rabson-Mendenhall syndrome
  - Lipoatrophic diabetes
  - Others
- Disease of the exocrine pancreas
  - Pancreatitis
  - Trauma/pancreatectomy
  - Neoplasis
  - Cystic fibrosis
  - Haemochromatosis
  - Fibrocalculou pancreatopathy
  - Others
- Endocrinopathies
  - Acromegaly
  - Cushing's syndrome
  - Glucagonoma
  - Phaeochromocytoma
  - Hyperthyroidism
  - Somatostatinoma
  - Aldosteronoma
  - Others
- Drug- or chemical-induced
  - Vacor
  - Pentamidine
  - Nicotinic acid
  - Glucocorticoids
  - Thyroid hormone
  - Diazoxide
  - β -adrenergic agonists
  - Thiazides
  - Dilantin
  - Interferon
    - γ -Interferon
    - α -Interferon
  - Others
- Infections
  - Congenital rubella
  - Cytomegalovirus
  - Others
- Uncommon forms of immune-mediated diabetes
  - "Stiff-man" syndrome
  - Anti-insulin receptor antibodies
  - Polyendocrine autoimmune deficiencies
    - APS I
    - APS II
  - Others
- Other genetic syndromes sometimes associated with diabetes
  - Down syndrome
  - Klinefelter syndrome
  - Turner syndrome
  - Wolfram syndrome
  - Friedreich's ataxia
  - Huntington's chorea
  - Laurence-Moon-Biedl syndrome
  - Myotonic dystrophy
  - Porphyria
  - Prader-Willi syndrome
  - Others

E15 Gestational diabetes mellitus

E12 Malnutrition-related diabetes mellitus

E14 Unspecified diabetes mellitus

E16 Impaired Glucose Regulation

- Impaired Glucome Tolerance (IGT)
- Impaired Fasting Glucose (IFG)

E17 Other disorders of glucose regulation and pancreatic internal secretion

- Nondiabetic hypoglycemia
  - Reactive hypoglycemia
  - Drug-induced (excluding insulin and oral hypoglycemic agents)
  - Critical organ failure
    - Hepatic diseases
    - Cardiac failure
    - Renal failure
  - Sepsis
  - Hormone deficiency
    - Cortisol
    - Growth hormone
    - Hypopituitarism
  - Insulin receptor antibody
  - Non-islet cell tumor
  - Insulin autoimmune syndrome
  - Factitious hypoglycemia
  - Insulinoma
  - Others
- Abnormal secretion of glucagon
  - Hyperplasia of pancreatic endocrine cells with glucagon excess
  - Glucagonoma
- Abnormal secretion of gastrin
  - Gastrinoma
  - Zollinger-Ellison syndrome
  - Drug-induced
- Disorder of pancreas internal secretion, unspecified

Disorders of amino-acid or proteins metabolism

Disorders of phenylalanine or tyrosine metabolism

- Phenylketonuria
  - Classical phenylketonuria — E70.0
  - BH4 deficiency
  - Classical phenylketonuria
    - Phenylalanine hydroxylase total deficiency
    - Phenylalanine hydroxylase partial deficiency
  - Atypical phenylketonuria
  - **Excludes: 6-pyruvoyl-tetrahydropterin synthase deficiency Include in neurology.**
- Maternal hyperphenylalaninaemia — E70.1
- Tyrosinaemia — E70.2
  - type 1
  - type 2 — E70.2
  - type 3 — E70.2
- Oculocutaneous tyrosinaemia
- Alkaptonuria — E70.2
- Ochronosis
- Hawkinsinuria
- Tyrosine oxidase temporary deficiency
- **Excludes: Albinism** — E70.3

Disorders of histidine metabolism — E70.8
- Histidinaemia
- Formiminoglutamic aciduria

Disorders of tryptophan or lysine metabolism — E70.8
- Hypertryptophanaemia
- Hyperlysinaemia
- Glutaryl-CoA dehydrogenase deficiency
- 2-aminoadipic aciduria
- Encephalopathy due to hydroxykynureninuria
- Seizures - intellectual deficit due to hydroxylysinuria
- Saccharopinuria
- Pellagra-like skin rash – neurological manifestations

Disorders of leucine, isoleucine or valine metabolism
- Leucinosis — E71.0
- Maple-syrup-urine disease
- 3-hydroxy-3-methylglutaric aciduria

Disorders of cystein or methionin metabolism — E72.1
- Cystathioninuria
- Classic homocystinuria
- Methioninaemia
- Sulfite oxidase deficiency
  - Isolated sulfite oxidase deficiency
  - Sulfite oxidase deficiency due to molybdenum cofactor deficiency
- N5-methylhomocysteine transferase deficiency
- **Excludes: Transcobalamin II deficiency** — D51.2

Disorders of ornithine or proline metabolism — E72.4
- Hyperornithinaemia
- Ornithinaemia
- Hyperprolinaemia type I
- Hyperprolinaemia type II

Disorders of serine or glycine metabolism — E72.5
- Non-ketotic hyperglycinaemia
  - Isolated nonketotic hyperglycinaemia type 1
  - Isolated nonketotic hyperglycinaemia type 2
- Sarcosinaemia
- Neurometabolic disorder due to serine deficiency
- 3-Phosphoserine phosphatase deficiency
- 3-Phosphoglycerate dehydrogenase deficiency

Disorders of amino-acid transport — E72.0
- Hartnup's disease
- Hyperdibasic aminoaciduria type 1
- Lysinuric protein intolerance
- Hyperdibasic aminoaciduria type 2
- Iminoglycinuria
- Excludes: Cystinuria

Organic acidurias
- Methylmalonic aciduria - homocystinuria
- Methylmalonic aciduria, vitamin B12 unresponsive
- Methylmalonic aciduria, vitamin B12 responsive
- Propionic aciduria
- Isovaleric aciduria
- Multiple carboxylase deficiency
- Ketoacidosis due to betaketothiolase deficiency
- 3 hydroxyisobutyric aciduria
- 3-hydroxy-3-methylglutaric aciduria
- 3-methylcrotonylglycinuria
- 3-methylglutaconic aciduria
- *MGA*
  - 3-methylglutaconic aciduria, type 1
  - 3-methylglutaconic aciduria, type 3
  - Costeff syndrome
  - 3-methylglutaconic aciduria, type 4
  - **Excludes: Barth syndrome (= 3-methylglutaconic aciduria, type 2)**
- 2-methylbutyryl-CoA dehydrogenase deficiency
- Isobutyryl-CoA dehydrogenase deficiency
- Glutaryl-CoA dehydrogenase deficiency
- 2-hydroxyglutaricaciduria
  - L-2-hydroxyglutaricaciduria
  - D-2-hydroxyglutaricaciduria
- Canavan disease
- Malonic aciduria
- Ethylmalonic aciduria encephalopathy
- **Excludes: 4-Hydroxybutyric aciduria**

Disorders of urea cycle metabolism — E72.2
- Argininaemia
  - Hyperargininaemia
  - Arginase deficiency
- Argininosuccinic aciduria
- Carbamoylphosphate synthetase deficiency
- Citrullinaemia
- Triple H (HHH) syndrome
- Hyperammonaemia due to N-acetylglutamate synthetase deficiency
- Progressive neurodegeneration - joint laxity - cataract
- Ornithine transcarbamylase deficiency

Hyperornithinemia — E72.4

Lysinuric protein intolerance

Gamma-glutamyl cycle disorder
- Glutathione synthetase deficiency
- 5-oxoprolinase deficiency
- Gamma-glutamyl transpeptidase deficiency
- Gamma-glutamylcysteine synthetase deficiency

Peptide metabolism disorder
- Carnosinaemia
- Prolidase deficiency
- Homocarnosinosis

Porphyria
- Porphyria, congenital erythropoietic — E80.0
- Protoporphyria, erythropoietic
- Porphyria, acute hepatic
  - Coproporphyria, hereditary
  - Porphyria, acute intermittent — E80.2
  - Porphyria variegata
  - Porphyria of Doss
- Porphyria, chronic hepatic
  - Hepatoerythropoietic porphyria
  - Porphyria cutanea tarda — E80.1

Other specified metabolic diseases by endogen intoxication
- French type sialuria
- Trimethylaminuria
- Lafora disease
- Hyperoxaluria

Energy metabolism disorder

Disorders of fatty-acid metabolism — E71.3
- Excludes: Schilder's disease — G37.0
- Muscle carnitine palmityltransferase deficiency
  - Carnitine palmitoyl transferase 1 deficiency
  - Carnitine palmitoyl transferase 2 deficiency
- Mitochondrial trifunctional protein deficiency
- Carnitine uptake deficiency
- Carnitine-acylcarnitine translocase deficiency
- Long chain 3-hydroxyacyl-CoA dehydrogenase deficiency
- Medium chain acyl-CoA dehydrogenase deficiency
- Multiple FAD dehydrogenase deficiency
- SCAD deficiency
- Acyl-CoA dehydrogenase, very long chain, deficiency of
  - Acyl-CoA dehydrogenase, long chain, deficiency of
  - Acyl-CoA dehydrogenase 9 deficiency
- 3-hydroxyacyl-CoA dehydrogenase, short chain, deficiency of
- 3-hydroxy 3-methylglutaryl-CoA (HMG) synthase deficiency

Mitochondrial respiratory chain disorders
- Kearns-Sayre syndrome
- MELAS syndrome
- MERRF syndrome
- NARP/MILS syndrome — H49.8
- Pearson syndrome
- Mitochondrial disease of nuclear origin
- NADH-CoQ reductase deficiency
- Cataract cardiomyopathy
- Leigh syndrome
  - Leigh syndrome due to cytochrome c oxidase deficiency
- Proximal tubulopathy - diabetes mellitus - cerebellar ataxia
- Lactic acidosis, congenital
- Barth syndrome
- Oxoglutaricaciduria
- Fumaric aciduria

Pyruvate dehydrogenase deficiency
Succinate CoQ reductase deficiency
Coenzyme Q cytochrome c reductase deficiency
ATP synthetase deficiency
Myoneurogastrointestinal encephalopathy syndrome
Coenzyme Q 10 (CoQ10), deficiency
Mitochondrial ADN deletions and duplications
Mitochondrial DNA depletion syndrome
GRACILE syndrome
Saguenay-Lac-St. Jean cytochrome oxidase deficiency
Ataxia - leukodystrophy - tubulopathy, due to cytochrome c oxidase deficiency
Cardiomyopathy - hypotonia, due to cytochrome c oxidase deficiency
Cardiomyopathy - hypotonia - lactic acidosis
Leukoencephalopathy with brain stem, spinal cord involvement - lactate elevation
Hypotonia with lactic acidaemia and hyperammonaemia
Deletion 2p21 (Hypotonia-cystinuria syndrome)
Fatal mitochondrial disease due to combined oxidative phosphorylation deficiency 3
Zellweger-like syndrome without peroxisomal anomalies
Alpers syndrome
Leber hereditary optic neuropathy
Wolfram syndrome
Excludes: Leber 'plus' disease
Excludes: Progressive external ophthalmoplegia

Creatine biosynthesis disorder
Guanidinoacetate methyltransferase deficiency
Arginine:glycine amidinotransferase deficiency
Intellectual deficit, X-linked, with seizures, short stature and midface hypoplasia

Carbohydrate metabolism disorder E74
Glycogen storage disease E74.0
Glycogen storage disease due to glycogen synthase deficiency (type 0)
Glycogen storage disease due to glycogen synthase deficiency, hepatic form
Glycogen storage disease due to glycogen synthase deficiency, muscular form
Glycogen storage disease due to glucose-6-phosphate system deficiency (type 1, Von Gierke disease)
Glycogen storage disease due to glucose-6-phosphatase deficiency (type 1A)
Glycogen storage disease due to glucose-6-phosphate translocase deficiency
Glycogen storage disease due to acid maltase deficiency (type 2 - Pompe disease)
Glycogen storage disease due to LAMP-2 deficiency (type 2B - Danon disease)
Glycogen storage disease due to glycogen debranching enzyme deficiency (type 3 - Cori-Forbes disease)
Glycogen storage disease due to glycogen branching enzyme deficiency (type 4 - Andersen disease - amylopectinosis)
Glycogen storage disease due to muscle phosphorylase deficiency (type 5 - McArdle disease)
Glycogen storage disease due to liver phosphorylase deficiency (type 6 or 6B - Hers disease)
Glycogen storage disease due to muscle phosphofructokinase deficiency (type 7 - Tarui disease)
Glycogen storage disease due to phosphorylase kinase deficiency (type 6A or 8 or 9)
Glycogen storage disease due to phosphoglycerate mutase deficiency (type 10 -Di Mauro disease)
Glycogen storage disease due to GLUT2 deficiency (type 11 - Bickel-Fanconi)
Glycogen storage disease due to aldolase A deficiency (type 12)
Glycogen storage disease due to enolase beta deficiency (type 13)
Glycogen storage disease due to phosphoglucomutase 1 deficiency (type 14)
Disorders of fructose metabolism E74.1
Essential fructosuria
Hereditary fructose intolerance
Disorders of galactose metabolism E74.2
Galactosaemia
Galactokinase deficiency
UDP-galactose-4-epimerase deficiency
Galactose-1-phosphate uridyltransferase deficiency
Gluconeogenesis disorder E74.4
Fructose-1,6-bisphosphatase deficiency
Phosphoenolpyruvate carboxykinase (PEPCK) deficiency
Phosphoenolpyruvate carboxykinase 1 (Pepck1) deficiency
Phosphoenolpyruvate carboxykinase 2 (Pepck2) deficiency
Pyruvate carboxylase deficiency

Ketolysis disorder
Ketoacidosis due to betaketothiolase deficiency
Succinyl-CoA acetoacetate transferase deficiency

Other specified energy metabolism disorders
Triose phosphate-isomerase deficiency
Multiple carboxylase deficiency
Multiple carboxylase deficiency, due to biotinidase deficiency
Multiple carboxylase deficiency, due to holocarboxylase synthetase deficiency
Phosphoglycerate kinase 1 deficiency
Succinic acidaemia
Glucosephosphate isomerase deficiency
Diphosphoglycerate mutase deficiency of erythrocyte
Dicarboxylicaminoaciduria
D-glycericacidaemia
Fatal infantile cytochrome C oxidase deficiency
Enolase deficiency
Lactate dehydrogenase deficiency
Phosphoglucomutase deficiency
Butyrylcholinesterase deficiency
Lipoamide dehydrogenase deficiency
Myoneurogastrointestinal encephalopathy syndrome
Glutaryl-CoA oxidase deficiency
Hyperinsulinism-hyperammonaemia syndrome
3-hydroxylacyl-CoA dehydrogenase deficiency
3-hydroxyacyl-CoA dehydrogenase, short chain, deficiency of
Excludes: Chondrodysplasia punctata lethal, neonatal
Excludes: Anaemia due to adenosine triphophatase deficiency
Excludes: Haemolytic anaemia due to red cell pyruvate kinase deficiency
Excludes: Anaemia due to adenosine triphosphatase deficiency

Defects in synthesis or catabolism of complex molecules
Peroxysomal disease
Refsum disease
Refsum disease, infantile form
Zellweger syndrome
Adrenoleukodystrophy, neonatal
Pseudoadrenoleukodystrophy
Pseudo-Zellweger syndrome
Bifunctional enzyme
deficiency
Acatalasaemia
Pipecolic acidaemia
Chondrodysplasia punctata, rhizomelic type
Bile acid synthesis defect, congenital, type 4

Lysosomal disease
Glycogen storage disease due to LAMP-2 deficiency
Neuronal ceroid lipofuscinosis
Congenital neuronal ceroid lipofuscinosis
Infantile neuronal ceroid lipofuscinosis
Late infantile neuronal ceroid lipofuscinosis
Juvenile neuronal ceroid lipofuscinosis
Adult neuronal ceroid lipofuscinosis
Progressive epilepsy-intellectual deficit, Finnish type
Lipid storage disease E75.5
Niemann-Pick disease, type C
Niemann-Pick C1 disease
Niemann-Pick C2 disease
Niemann-Pick disease, Nova Scotia type
Niemann-Pick D disease
Wolman disease
Cholesteryl ester storage disease
Lysosomal transport defect
Free sialic acid storage disease
Cystinosis

Mucopolysaccharidosis E76
Mucopolysaccharidosis type 1 E76.0
Hurler syndrome
Scheie syndrome
Hurler-Scheie syndrome
Mucopolysaccharidosis type 2 (Hunter syndrome) E76.1
Mucopolysaccharidosis type 3 (Sanfilippo syndrome) E76.2
Mucopolysaccharidosis, type 3A (Sanfilippo syndrome, type A)
Mucopolysaccharidosis, type 3B (Sanfilippo syndrome, type B)
Mucopolysaccharidosis, type 3C (Sanfilippo syndrome, type C)
Mucopolysaccharidosis, type 3D (Sanfilippo syndrome, type D)
Mucopolysaccharidosis type 4 (Morquio syndrome) E76.2
Mucopolysaccharidosis type 6 (Maroteaux-Lamy syndrome) E76.2
Mucopolysaccharidosis type 7 (Sly syndrome) E76.2
Mucopolysaccharidosis type 9 (Natowicz syndrome)
Oligosaccharidosis E77.1
Alpha-mannosidosis
Aspartylglucosaminuria
Beta-mannosidosis
Fucosidosis
Sialidosis
Lipomucopolysaccharidosis
Mucolipidosis type 1
Sialidosis type 1
Sialidosis, type 2
N-acetyl-alpha-D-galactosaminidase deficiency
Galactosialidosis
Mucolipidosis E77.0
Mucolipidosis type 2
Mucolipidosis type 3
Mucolipidosis type 4
Excludes: Sialidosis (mucolipidosis type 1)
Sphingolipidosis E75
Farber lipogranulomatosis
Krabbe disease
Fabry disease

- Metachromatic leukodystrophy
  - late infant type
  - Juvenile type
  - Adult type
  - Multiple sulfatase deficiendy
- GM1 gangliosidosis
- GM2 gangliosidosis
  - Tay-Sachs
  - Juvenile type
  - Adult type
  - Sandhoff disease
- Gaucher disease
  - type 1
  - type 2
  - type 3
  - Perinatal-lethal type
- Niemann-Pick disease
  - type A
  - type B
- Saposin deficiency

Protein glycosylation disorder

- CDG syndrome
  - **Excludes : CDG syndrome, type IIc (Leukocyte adhesion deficiency type 2)**

Sterol metabolism disorder

- Bile acid disorder
  - Cerebrotendinous Xanthomatosis
  - Excludes : Bile acid synthesis defect with cholestasis and malabsorption Include in hepatology.
    - Bile acid synthesis defect, congenital, type 1
    - Bile acid synthesis defect, congenital, type 3
    - Bile acid synthesis defect, congenital, type 2
- Sterol biosynthesis disorder
  - Mevalonicaciduria
  - Desmosterolosis
  - Lathosterolosis
  - CHILD syndrome
  - Excludes: Antley-Bixler syndrome
  - Excludes: Greenberg dysplasia
  - Excludes: Smith-Lemli-Opitz syndrome
  - Excludes: X-linked dominant chondrodysplasia punctata
  - Excludes: Partial mevalonate kinase deficiency with recurrent fever +/- hyperIgD
  - Excludes: Hyperimmunoglobinaemia D with recurrent fever

Purine or pyrimidine metabolism disorder

- Purine metabolism disorder
  - Lesch-Nyhan syndrome — E79
    - Kelley-Seegmiller syndrome
  - Adenosine monophosphate deaminase deficiency
  - Purine nucleoside phosphorylase deficiency
  - Adenylosuccinate lyase deficiency
  - Phosphoribosylpyrophosphate synthetase superactivity
  - Xanthinuria
  - Excludes: 2,8 dihydroxyadenine urolithiasis
- Pyrimidine metabolism disorder
  - Orotic aciduria hereditary
  - Dihydropyrimidine dehydrogenase deficiency
  - Myoneurogastrointestinal encephalopathy syndrome
  - Dihydropyrimidinuria
  - Beta-ureidopropionase deficiency
  - Excludes: Anaemia due to pyrimidine 5' nucleotidase deficiency

Disorders of mineral metabolism — E83

Excludes: Dietary mineral deficiency — E58-E61

Excludes: Parathyroid disorders — E20-E21

Excludes: Vitamin D deficiency — E55

Excludes: Disorders of iron metabolism, including:

- Iron overload disease
  - Haemochromatosis — E83.1
  - Ferroportin disease
  - Neonatal haemochromatosis
  - African iron overload
  - Friedreich ataxia
  - Neurodegeneration with brain iron accumulation
  - Aceruloplasminaemia
  - Atransferrinaemia
  - Microcytic anaemia with liver iron overload
- Iron deficiency anaemias — D50
- Sideroblastic anaemias — D64.0-D64.3

Excludes: Non-endocrine disorders of phosphocalcic metabolism are

- Non-endocrine disorders of phosphorus metabolism, including: — E83.3
  - Familial hypophosphataemia
  - Hypophosphatasia
  - Vitamin-D-resistant osteomalacia
  - Vitamin-D-resistant rickets — M83
  - adult osteomalacia
  - osteoporosis — M80-M81
- Non-endocrine disorders of calcium metabolism — E83.5
  - Familial hypocalciuric hypercalcaemia
  - Idiopathic hypercalciuria
  - chondrocalcinosis — M11.1-M11.2
  - adult osteomalacia — M83
  - osteoporosis — M80-M81

Disorders of copper metabolism — E83.0

- Menkes disease
- Kinky hair disease
- Steely hair disease
- Occipital horn syndrome
- Wilson disease
- Benign familial copper deficiency

Disorders of zinc metabolism — E83.2

- Acrodermatitis enteropathica

Disorders of magnesium metabolism — E83.4

- Hypermagnesaemia
- Hypomagnesaemia

Other specified disorders of mineral metabolism — E83.8

Disorders of lipoprotein metabolism and other lipidaemias (dyslipidaemia) — E78

- Hyperlipidaemia
  - Hypercholestorolaemia (high LDL cholesterol (type IIa hyperlipoproteinaemia))
    - primary hypercholesterolaemia
      - familial hypercholesterolaemia: FH
        - FH homozygote
        - FH heterozygote
        - familial detective apo B—100
        - autosomal recessive hypercholesterolaemia
        - polygenic hypercholesterolaemia
    - secondary hypercholesterolaemia
  - Hypertriglyceridemia
    - type I hyperlipoproteinemia
      - familial chylomicron syndrome
      - familial lipoprotein lipase (LPL) deficiency
      - apolipoprotein C-II deficiency
    - type IV hyperlipoproteinaemia
      - familial typeIV hyperlipoproteinemia
      - secondary typeIV hyperlipoproteinemia
    - type V hyperlipoproteinaemia
      - familial typeV hyperlipoproteinemia
      - secondary typeV hyperlipoproteinemia
  - Combined hyperlipidaemia — E78.2
    - type IIb hyperlipoproteinaemia
      - familial combined hyperlipidaemia
      - secondary combined hyperlipidaemia
    - type III hyperlipoproteinaemia
      - familial dyslipoproteinaemia
      - familial detective apoE
      - familial apoE deficiency
      - secondary type III hyperlipoproteinaemia
- Hypolipoproteinemia
  - hypoalphalipoproteinemia (low HDL cholesterol)
    - familial hypoalphalipoproteinemia
      - Tangier disease
      - apolipoprotein A-I deficiency
      - familial LCAT deficiency
      - fish—eye disease
    - secondary hypoalphalipoproteinemia
  - hypolipoproteinaemia
    - Abetalipoproteinaemia
    - Microsomal triglyceride transfer protein (MTP) deficiency
    - familiar hypobetalipoproteinaemia
    - Truncated form of apolipoprotein B-100
    - secondary hypobetalipoproteinaemia
  - Other dyslipidaemia
    - primary hyperalphalipoproteinemia (high HDL cholesterol)
      - Cholestery ester transfer protein (CETP) deficiency
      - Hepatic triglyceride lipase (HTGL) deficiency
    - Sitosterolaemia

Disorders of fluid, electrolyte and acid-base balance

Other specified rare metabolic disorders — E88.8

消化器分野

Proposed new GI code plan after ICD-11 alpha-draft version by Gastroenterology (GI) Working Group of IM-TAG.

–Revised Nov 2010 K-FLAG PLAN-

(1) Oral cavity

(2) Diseases of oesophagus (KD)

KDBA Congenital anomalies of the oesophagus
(congenital malformations of oesophagus (Q39))
(excludes: hiatal hernia K90)
* 0. Atresia of oesophagus without fistula (Q39.0)
* 1. Atresia of oesophagus with tracheo-oesophageal fistula (Q39.1)
* 2. Congenital tracheo-oesophageal fistula without atresia (Q39.2)
* 3. Congenital stenosis and stricture of oesophagus (Q39.3)
* 4. Esophageal congenital web (Q39.4)
* 5 . Congenital dilatation of oesophagus (Q39.5)
Congenital cardiospasm
*6. Diverticulum of oesophagus (congenital)(Q39.6)
Esophageal pouch (congenital)
*7. Heterotopic gastric mucosa (Inlet patch)
*8. Other specified congenital anomalies of oesophagus (Q39.9)
Absence
Congenial displacement
Esophageal Duplication
Congenital duplication cyst

KDBB Anatomical alterations of the oesophagus
(excludes: congenital anomalies)
KDBA. Oesophageal obstruction due to compression, stricture or stenosis
(excludes: due to GERD K19, congenital anomalies K16.0, oesophageal cancer K25, foreign body K17.8)
(Use additional cause code *(Chapter XX), if desired, to identify cause)
due to chemical agents
etc.
KDBB. Diverticulum of oesophagus (acquired)

Oesophageal pouch, acquired
Zenkers diverticulum
KDBC. Oesophageal web
(includes:
*Sideropenic dysphagia (D50.8),
*Plummer-Vinson syndrome, or
*Paterson-Kelly syndrome)
KDBD. Megaoesophagus
Idiopathic megaoesophagus (excludes: achalasia K18.0)
* in Chagas disease (B57.3)
KDBE. Fistula of oesophagus (excludes: congenital fistula, malignant neoplasm)
KDBF. Perforation/rupture of oesophagus
(excludes: Mallory-Weiss syndrome, malignant neoplasm)
0. Boerhaave syndrome (idiopathic)
Use additional code if desired, to identify cause.
*1. injury/trauma
*2. due to endoscopic procedures
KDBG. Gastric heterotopia of oesophagus
KDBH. Other and unspecified anatomical alterations of the oesophagus

KDC Motility disorders of oesophagus
(Dyskinesia of oesophagus K22.4)
KDCA. Achalasia (K22.0)
Cardiospasm, Non-Chagas
(excludes: congenital cardiospasm K16.05, Q39.5)
*Achalasia- Chagas (B57.3)
KDCB. Motility disorders of cervical and upper esophagus
KDCC. Disorder of esophageal peristalsis
(excludes: achalasia, GERD)
0. Hypertensive peristalsis
Spasm of oesophagus
Diffuse oesophageal spasm
Corkscrew oesophagus
1. Spastic peristalsis
Nutcracker oesophagus
2. Hypotensive peristalsis
KDCD. Disorders of lower oesophageal sphincter function
0. Hypertensive LES
1. Hypotensive LES
2. Incomplete LES relaxation
*KDCE. Oesophageal dysmotility associated with systemic diseases

(Use additional cause code to identify cause.)
KDCF. Other specified or unspecified motility disorders of oesophagus

KDD Gastro-oesophageal reflux disease (GERD/GORD)(K21)
KDDA. Non-erosive reflux disease (NERD)
GERD/GORD without oesophagitis or erosion, with or without hiatus hernia.
(Excludes: functional heartburn)
KDDB. Erosive oesophagitis
(incl: Oesophagitis: peptic)
GERD/GORD with oesophagitis and/or erosion, with or without hiatus hernia (K21.0).
Reflux oesophagitis
KDDC. Erosive oesophagitis with complications
with stenosis
hemorrhage
anemia (occult bleeding)
KDDD. Ulcer of oesophagus due to GERD/GORD
(Ulcer of oesophagus: peptic)
KDDE. Ulcer of oesophagus due to GERD/GORD with complication
with perforation
stenosis or obstruction
hemorrhage
anemia (occult bleeding)
KDDF. Other and unspecified GERD

KDE Columnar metaplastic epithelium in the oesophagus (Barrett's oesophagus K22.7)
KDEA. Barrett's epithelium
00. gastric metaplasia
01. intestinal metaplasia
KDEB. Dysplasia of Barrett's epithelium
-low grade
-high grade
KDEC. Barrett's ulcer
KDED. *Barrett's adenocarcinoma (see Barrett's cancer, Oesophageal adenocarcinoma K25)
KDEE. Other and unspecified Barrett's oesophagus

KDF Oesophagitis
(excludes: reflux oesophagitis K19, oesophageal ulcer/erosion K22,

Crohn's disease K52)
KDFA. Infectious oesophagitis coded elsewhere
(Code also infectious code to specify causative organisms)
0. Bacterial
(incl. Oesophageal phlegmone, Abscess of oesophagus)
1. Viral
2. Fungal
3. Parasitic
KDFB. Eosinophilic oesophagitis
KDFC. Oesophagitis due to systemic disorders classified elsewhere
* Sarcoidosis
* Collagen diseases
* Dermatological diseases (e.g. Epidermolysis bullosa)
(Use additional disease code to identify cause.)
KDFD. Oesophagitis due to external causes
0. Drug-induced esophagitis
Use external cause code *(Chapter XX) to identify cause.
1. Chemical oesophagitis (Oesophagitis: chemical)
Use external cause code *(Chapter XX) to identify cause.
2. Radiation/Thermal oesophagitis
3. Other external causes
(Use external cause code *(Chapter XX), if desired, to identify cause.)
foreign body
due to endoscopic procedures
sequelae of endoscopic therapy, etc.
KDFE. Other or unspecified oesophagitis
Oesophagitis not elsewhere classified

KDF Oesophageal ulcer and erosion
(includes: Ulcer of oesophagus (K22.1), Ulcerative oesophagitis)
(excludes: reflux disease, Barrett's ulcer, malignant esophageal ulcer, Crohn's disease K52)
KDFA. Oesophageal erosion
(Erosion of oesophagus)
KDFB. Infectious oesophageal ulcer coded elsewhere
(Go to infectious code to specify causative organisms)
Bacterial
Viral
Fungal
(Ulcer of oesophagus: fungal)

Parasitic

KDFC. Oesophageal ulcer due to allergic/immunologic disorders
(excludes: Crohn's disease K52)
Eosinophilic/allergic
Collagen diseases
etc.

KDFD. Oesophageal ulcer due to external causes
0. Drug-induced esophageal ulcer
(Ulcer of oesophagus: due to ingestion of drugs and medicaments)
(Use external cause code *(Chapter XX), if desired, to identify cause.)
1. Chemical esophageal ulcer
(Ulcer of oesophagus: due to ingestion of chemicals)
(Use external cause code *(Chapter XX), if desired, to identify cause.)
2. Radiation/thermal esophageal ulcer
Radiation
Thermal
3. Other external causes
(Use additional cause code, if desired, to identify cause.)
injury/trauma (S27.8)
foreign body (T18.1)
prosthetic device
due to endoscopic procedures
sequelae of endoscopic therapy, etc.

KDFE. Oesophageal ulcer, etiology unspecified with complication
with perforation
with obstruction or stenosis
with hemorrhage
with anemia (with occult bleeding)

KDFF. Other and unspecified oesophageal ulcer
Ulcer of oesophagus: NOS

KDH Oesophageal varices (K20, H)

(link to portal hypertension in liver section, includes: Intramucosal venous dilatation of oesophagus (IMVD), Oesophageal varices without bleeding in disease classified elsewhere K20.6, Oesophageal varices with bleeding in disease classified elsewhere K20.7, Oesophageal varices in: liver disorders, Osophageal varices in: schistosomiasis)

KDHA. Oesophageal varices without bleeding

Oesophageal varices NOS
Osophageal varices, unspecified

KDHB. Oesophageal varices with bleeding

KDHC. Oesophageal non-bleeding varices with gastric varices
0. with non-bleeding gastric varices
1. with bleeding gastric varices

KDHD. Oesophageal bleeding varices with gastric varices
0. with non-bleeding gastric varices
1. with bleeding gastric varices

KDHE. Downhill varices of the oesophagus (T)

KDHF. Solitary varices of the oesophagus

KDHG. Other specified and unspecified oesophageal varices
0. Intramucosal venous dilatation of oesophagus (IMVD)

KDI Other vascular disorders of oesophagus
(excludes: esophageal varices KDH)

KDIA. Angiodysplasia and arteriovenous malformation (AVM) of the oesphagus

KDIB. Hemangioma (Please see K27 Other neoplasm of esophagus).

KDIC. Intramural haemorrhage of oesophagus
(excludes: esophageal varices K23)
(Use additional code if desired, to identify cause)
* injury/trauma
* due to endoscopic procedures etc.

KDID. Gastro-esophageal lesion (laceration)-hemorrhage syndrome
Mallory-Weiss syndrome
Mallory-Weiss lesion
Mallory-Weiss tear

KDIE. Other and unspecified vascular disorders of esophagus
Oesophageal haemorrhage unknown origin

KDJ Neoplasm of the oesophagus

***KDJA Malignant neoplasm of oesophagus**

*0. Oesophageal cancer
(excludes: oesophageal juctional cancer KDJA1)
(includes: oesophageal cancer with complication, eg. hemorrhage, obstruction/stenosis, perforation)
*00. Squamous cell cancer (SCC)
*01. Adenocarcinoma
(Incl. Barrett's cancer)
09. Other specified and unspecified esophageal cancer
adenoid cystic, adenosquamous, mucoepidermoid carcimona, undiffirentiated, etc.

1. Oesophageal junctional cancer
   10 Adenocarcinoma (should be subdivided in AEG 1,AEG 2)
      AEG 3 see gastric cancer proximal (subcardial) location
      (Incl. Barrett's cancer)
   19 Other and unspecified oesophageal junctional cancer
2. Other primary malignant neoplasm of the oesophagus
   (excludes: esophageal cancer K25 and K26, and Barrett's related cancer K20.3)
   20. Malignant mesenchymal tumours
      200. Leiomyosarcoma
      201. Malignant GIST
      202. Other malignant mesenchymal tumours
         (Hemangiomasarcoma, melanoma, etc.)
   21. Neuroendocrine neoplasm
      210. Neuroendorine tumor (NET)(incl. carcinoid)
      211. Neuroendocrine carcinoma (NEC)
      212. Other neuroendocrine neoplasm
   22. Lymphomas

*3. Metastatic or secondary malignant neoplasm of oesophagus
*9. Other and unspecified malignant neoplasm of oesophagus

KDJB. Premalignant lesions
(incl. Carcinoma in situ)
Intraepithelial neoplasia (severe dysplasia)
Dysplasia (low grade, high grade)
(Includes Barrett's dysplasia)

KDJC. Benign neoplasm
* Benign epithelial tumor (incl. hyperplastic polyp)
* Squamous cell papilloma
*Granular cell tumor
*Hemangioma
*Leiomyoma
*GIST (non-malignant)
*Other mesenchymal tumors

KDJD. Other specified and unspecified neoplasm of the oesophagus
Neoplasm of uncertain or unknown behaviour

KDJE. Oesophageaal neoplasm, histology unspecified, with complication
(This code could be used in combination with other code in KDJ as double code, if desired)
0. with hemorrhage

1. with anemia (occult blood)
2. with fistula
3. with perforation
4. with obstruction/stenosis

KDJF. Oesophageal neoplasm, histology unspecified, located at
(location code could be used in combination with other code in KDJ as double code, if desired)
50.Upper third of oesophagus
Cervical part of oesophagus
51. Middle third of osepophagus
Thoracic part of oesophagus
52. Lower third of oesophagus
Abdominal part of oesophagus

KDK Other diseases of the oesophagus
0. Foreign body with/without complications
60. without complication
61. with complication
hemorrhage
obstruction
perforation
8. Other specified diseases of oesophagus (K22.8)
(incl: Disorders of oesophagus in other diseases classified elsewhere (K23.8))
9. Disease of oesophagus, unspecified (K22.9)

(3) Diseases of Stomach (KE)

(excludes: Hiatal hernia K90, Disorders of gastrostomy K98,
Esophageal junctional cancer (except: adenocarcinoma of the esophagogastric junction:AEG) 3 K26)

KEA Congenital anomalies of the stomach

*KEAA. Congenital hypertrophic pyloric stenosis (Q40.0)
*KEAB. Aberrant pancreas in the stomach
*KEAC. Other specified congenital anomalies of the stomach
*Gastric atresia (Q40.2)
*Microgastria
* Gastric duplication (Q40.2)
*Congenital diverticulum of stomach
*Congenital gastric malrotation
*Displacement
*Megalogastria
*Congenital hourglass stomach
*KEAD. Unspecified congenital anomalies of the stomach

KEB Anatomical alterations of the stomach

(excludes: congenital anomalies)
KEBA. Gastric outlet obstruction due to compression, stricture, or stenosis)
(includes: Adult hypertrophic pyloric stenosis)
(excludes: functional obstruction, congenital anomalies (K24.01), due to ulcer, due to neoplasm)
0. Adult hypertrophic pyloric stenosis
Pyloric stenosis NOS
1. Gastric stenosis due to other causes
(Use additional code, if desired, to identify cause)
KEBB. Gastric fistula
KEBC. Gastric volvulus
KEBD. Perforation of stomach
Use external cause code *(Chapter XX), if desired, to identify cause.
injury/trauma
due to endoscopic procedures
etc.
KEBE. Gastric diverticulum
(excludes: congenital diverticulum of stomach)
KEBF. Other specified and unspecified structural alteration of stomach
Hourglass stricture and stenosis of stomach (K31.2)
(excludes: congenital hourglass stomach

hourglass contraction of stomach)

KEC Gastroduodenal motility and secretory disorders

KECA. Dumping syndrome (includes: post-gastrectomy dumping Syndrome, postvagotomy syndrome)
KECB. Abnormal gastric motility
Gastroparesis (link to diabetes)
Other motility disorder
KECC. Acute dilatation of stomach (K31.0)
Acute distension of stomach
KECD. Acid hypersecretion (excludes: Zollinger-Ellison syndrome K82.12, E16.4)
eg. mastocytosis
KECE. Achlorhydria
KECF. Other specified and unspecified motor and secretory disorders

KED Gastritis

(Inflammation of gastric mucosa, refer to dyspepsia or functional dyspepsia for symptomatic conditions)
(includes: Gastritis with specific endoscopic or pathologic features, eg. Atrophic gastritis (Gastric atrophy), Metaplastic gastritis (Intestinal metaplasia), Acute hemorrhagic gastritis, Acute erosive and hemorrhagic gastric lesions (AGML-link to the section on ulcer/erosions)

KEDA. H.pylori-induced
KEDB. Chemical gastritis
0. Alcoholic gastritis (K29.1)
Inflammation of the gastric mucosa due to alcoholic use
1. Drugs and medicaments
2. Biliary reflux
KEDC. Autoimmune gastritis
KEDD. Other specific causes of gastritis
Eosinophilic
Allergic and dietetic
Lymphocytic
Ménétrier disease
KEDE. Infectious gastritis (coded elsewhere)
Bacterial
(except. Helicobacter pylori-induced)
(incl. gastric phlegmone, gastric abscess)
Viral

Fungal
Parasitic
KEDF. Due to systemic disorders classified elsewhere
0. Sarcoidosis
1. Vasculitis
(excludes: Crohn's disease K52)
KEDG. Due to other external causes
(Use external cause code *(Chapter XX), if desired, to identify cause.)
0. radiation/thermal
1. foreign body
2. due to endoscopic procedures
etc
KEDH. Gastritis, etiology unspecified, with specific endoscopic or pathologic features
0. Superficial gastritis
00. Acute gastritis
Other acute gastritis (K29.1)
01. Chronic superficial gastritis (K29.3)
1. Acute haemorrhagic gastritis (K29.0)
Acute (erosive) gastritis with hemorrhage
(incl. Acute gastric mucosal lesion (AGML))
(excludes: gastric erosion- mainly chronic)
2. Chronic atrophic gastritis (K29.4)
(Gastric atrophy, Chronic gastritis, unspecified (K29.5))
20. Mild to moderate atrophy
21. Severe gastric atrophy
3. Metaplastic gastritis (Intestinal metaplasia)
4. Granulomatous gastritis (NOS)
5. Hypertrophic gastritis (excludes :Ménétrier disease K31.3)
KEDG. Other and unspecified gastritis
(Other gastritis K29.6)
0. Gastroudodenitis (K.29.9)
Gastritis and duodenitis
1. Gastroenteritis
Gastroenteritis and colitis
9. Gastritis, unspecified K29.7)

KEE Gastric ulcer and erosion
(includes: Peptic ulcer disease in the stomach, mucosal defect, ulcer scar, Zollinger-Ellison syndrome, gastric erosions)

(excludes: malignant ulcer, Dieulafoy ulcer K29.52)
KEEA. Gastric erosion (excludes: AGML, acute erosive gastritis KEDH1)
KEEB. Gastric ulcer- Helicobacter-pylori-induced
KEEC. Gastric ulcer drug/toxic-related
(includes NSAIDs ulcer)
(Use additional code if desired, to identify cause)
KEED. Gastric ulcer due to both Helicobacter pylori and drugs
KEEE. Other specific causes of gastric ulcer
(excludes: Crohn's disease K52)
0. *Zollinger-Ellison syndrome
1. Eosinophilic
2. Lymphocytic
3.*Infectious diseases coded elsewhere
Bacterial
Viral
Fungal
Parasitic
4. Due to systemic diseases classified esewhere
Sarcoidosis
Vasculitis
5. Stress ulcer and ulcerations (intensive care related)
6. Other external causes
Iatrogenic
(Use external cause code *(Chapter XX), if desired, to identify cause)
KEEF. Gastric ulcer etiology unspecified, without complication (idiopathic)
0. Acute gastric ulcer
(includes. Gastric ulcer: acute without haemorrhage or perforation K25.3)
1. Chronic gastric ulcer
(includes. Gastric ulcer: chronic without haemorrhage or perforation K25.7)
2. Gastric ulcer: unspecified as acute or chronic, without haemorrhge or perforation K26.9)
KEEG. Gastric ulcer etiology unspecified, with complication
0. with hemorrhage
00. acute with haemorrhage (K25.0)
01. chronic or unspecified with haemorrhage (K25.4)
1. with anemia (with occult bleeding)
2. with perforation
20. acute with perforation (K25.1)

21. acute with both perforation and haemorrhage (K25.2)
22. chronic or unspecified with perforation (K25.5)
23. chronic or unspecified with both perforation and haemorrhage (K25.6)
3. with obstruction or stenosis

KEEH. Gastric ulcer etiology unspecified, located at
(location code could be used in combination with other codes in KEE as double code, if desired)
*0 proximal region including fornix and cardia
*1 middle region including angle
*2 distal region including antrum and pylorus
*3 In the remnant stomach (not the anastomatic K28)

KEEI. Other and unspecified peptic ulcer
0.Peptic ulcer, site unspecified, etiology unspecified (K27).
01.Acute peptic ulcer, site unepcified
(includes. Peptic ulcer, site unspecified : acute without haemorrhage or perforation K27.3)
02. Chronic peptic ulcer, site unspecified
(includes.Peptic ulcer, site unspecified: chronic without haemorrhage or perforation K27.7)
03. Peptic ulcer, site unspecified, unspecified as acute or chronic, without haemorrhge or perforation K27.9)
1. Peptic ulcer, site unspecified, etiology unspecified, with complication
10. with hemorrhage
100. acute with haemorrhage (K27.0)
101. chronic or unspecified with haemorrhage (K27.4)
11. with anemia (with occult bleeding)
12. with perforation
120. acute with perforation (K27.1)
121. acute with both perforation and haemorrhage (K27.2)
122. chronic or unspecified with perforation (K27.5)
123. chronic or unspecified with both perforation and haemorrhage (K27.6)
13. with obstruction or stenosis

KEF Anastomotic ulcer and erosion of upper gastrointestinal tract
(includes: Peptic ulcer disease of anastomotic sites, erosion)
(includes: After esophagogastric, gastroduodenal, gastrojejunal, gastrointestinal, gastrocolic, anastomosis)
KEFA. Anastomotic erosion
KEFB. Anastomotic ulcer, peptic
KEFC. Anastomotic ulcer, HP-induced

KEFD. Anastomotic ulcer drug-induced
KEFE. Anastomotic ulcer other causes
KEFF. Anastomotic ulcer etiology unspecified, without complication
0. Acute anastomotic ulcer
(includes. Gastrojejunal ulcer: acute without haemorrhage or perforation K28.3)
1. Chronic anastomotic ulcer
(includes. Gastricjejunal: chronic without haemorrhage or perforation K28.7)
2. Anastomotic ulcer: unspecified as acute or chronic, without haemorrhge or perforation K28.9)

KEFG. Anastomotic ulcer etiology unspecified, with complication
0. with hemorrhage
00. acute with haemorrhage (K28.0)
01. chronic or unspecified with haemorrhage (K28.4)
1. with anemia (with occult bleeding)
2. with perforation
20. acute with perforation (K28.1)
21. acute with both perforation and haemorrhage (28.2)
22. chronic or unspecified with perforation (K28.5)
23. chronic or unspecified with both perforation and haemorrhage (K28.6)
3. with obstruction or stenosis

KEFH. Anastomotic ulcer etiology unspecified, located at
(location code could be used in combination with other codes in KEE as double code, if desired)
0. esophagogastric,
1. gastroduodenal,
2. gastrointestinal
gastrojejunal K28
3. gastrocolic,
4. other anastomotic (marginal)

KEFI. Other and unspecified anastomotic ulcer and erosion

KEG Gastric varices
(without oesophageal varices, please refer oesophageal varices if oesophageal varices are complicated)
0. Gastric varices non-bleeding
1. Gastric varices with bleeding

KEH Other vascular disorders of the stomach
(excludes: gastric varices K34)
KEHA. Angiodysplasia and arteriovenous malformation (AVM)

0. anigodysplasia
includes: Hereditary hemorrhagic teleangiectasia
(Rendu-Osler Weber disease)(I78.0)
1. Dieulafoy lesion
2. Other A-V malformation

KEHB *Hemangioma (Please see K39 Other Neoplasm of stomach)
Capillary hemangioma
Cavernous hemangioma

KEHC. Portal hypertensive gastropathy

KEHD. Vascular ectasia
(includes: GAVE, Water melon stomach)

KEHE. Intramural haemorrhage of the stomach
*due to injury/trauma
(Use external cause code *(Chapter XX), if desired,
to identify cause.)
other causes

KEHF. Other and unspecified vascular disorders of the stomach
Gastric haemorrhage unknown origin

KEG Gastric polyps
(excludes: adenomatous polyp, adenoma KEHB)

KEGA. Non-neoplasitc polyps
0. Hyperplastic polyp
1. Fundic 'cystic' gland polyp

KEGB. Gastric polyps, histology unspecified, located at
(location code could be used in combination with other
etiology code in KEG as double code, if desired)
0. Proximal
1. Middle
2. Distal
3. In the remnant stomach (not the anastomatic K28)
4. Other and unspecified
Overlapping lesion of stomach
Both stomach and duodenum (K31.7)
Unspecified

KEGC. Other and unspecified gastric polyps

KEH Neoplasms of the stomach
(excludes: gastric polyps KEG)

KEHA Malignant neoplasm of stomach

*0. Gastric cancer (C16)
(primary, excludes: secondary gastric cancer)
00. Adenocarcinoma

01. Other histologic type
adenosquamous, medullary, hepatoid, squamous cell,
undifferentiated etc.)

1. Gastric lymphoma
*10. MALT-type lymphoma
100. H.pylori related-related (C88.4)
101. MALT-type lymphoma etiology unknown (C88.4)
11. Diffuse non-Hodgkin's lymphoma
12. Other specified and unspecified gastric lymphoma
Follicular non-Hodgkin's lymphoma
Mantle cell lymphoma

2. Other primary malignant neoplasm of the stomach
20. Malignant mesenchymal tumours
200. Leiomyosarcoma
201. Malignant GIST
202. Other malignant mesenchymal tumours
(Kaposi sarcoma, synovial sarcoma, etc.)
21. Neuroendocrine neoplasm
210. Neuroendorine tumor (NET)(incl. carcinoid)
211. Neuroendocrine carcinoma (NEC)
212. Other neuroendocrine neoplasm
Gastrin-producing NET, Serotonin-producing NET etc.

3.. Metastatic or secondary malignant neoplasm of the stomach

KEHB. Premalignant lesions
(incl. Carcinoma in situ, adenoma)
0. Adenomatous polyp (Adenoma)
1. Intraepithelial neoplasia
(incl. severe or high grade dysplasia)
2. Dysplasia (low grade)

KEHC. Benign neoplasm other than gastric polyps
*Hemangioma
*Leiomyoma
*GIST (non-malignant)
*Other benign neoplasm
Inflammatory myofibroblastic tumour, Glomus tumour,
Plexiform fibromyxoma etc.

KEHD. Other specified and unspecified neoplasm of the stomach
Neoplasm of uncertain or unknown behaviour

KEHE. Gastric neoplasm, histology unspecified, with complication
(This code could be used in combination with other code in

KEH as double code, if desired)
Malignant neoplasm, histology unspecified,
0. with hemorrhage
1. with anemia (occult blood)
2. with perforation
3. with obstruction/stenosis

KEHF. Gastric neoplasm, histology unspecified, located at
(location code could be used in combination with other etiology code in KEH as double code, if desired)
0. Proximal (includes AEG 3)
(excludes: oesophagogastric junctional cancer K26-AEG 1, AEG 2)
Cardia
Fundus
1. Middle including angle
Body of stomach
2. Distal
Pyloric antrum
Pylorus
3. In the remnant stomach (not the anastomatic K28)
4. Other and unspecified
Overlapping lesion of stomach
Lesser curvature of stomach, unspecified
Greater curvature of stomach, unspecified
unspecified

KEI Other diseases of the stomach
KEIA. Foreign body in the stomach
0. without complication
eg. Coin
Gastric bezoars (T18.2)
1. with complication
hemorrhage
obstruction
perforation

(4) Diseases of duodenum (KF)

KFA Congenital anomalies of the duodenum
KFAA. *Congenital absence, atresia and stenosis of duodenum
KFAB * Duodenal duplication
KFAC Gastric heterotopia in the duodenum
*KFAD Other and unspecified congenital anomalies
0. Congenital intraduodenal diverticulum
*1. Megaduodenum

KFB Anatomical alterations of the duodenum
(excludes: congenital anomalies)
KFBA. Obstruction of duodenum due to compression, stricture, or stenosis
(excludes: congenital stenosis K32.00)
Duodenal ileus, Stenosis of duodenum
(excludes: duodenal ulcer, duodenal neoplasm)
Use additional code, if desired, to identify cause.
*0. postsurgical
*1. inflammatory
KFBB. Diverticulum of duodenum (acquired)
KFBC. Duodenal fistula
(due to cholelithasis etc.)
KFBD. Perforation of duodenum
(excludes; duodenal ulcer, duodenal neoplasm)
(Use external cause code *(Chapter XX), if desired, to identify cause.)
*0 injury/trauma
*1 due to endoscopic procedures
*2 due to other medical procedures (vascular coiling etc)
etc.
KFBE. Deformity of duodenum
Use additional code, if desired, to identify cause.
KFBF. Other and unspecified anatomical alterations of the duodenum

KFC Duodenitis
(includes: duodenal inflammation)
(excludes: Crohn's disease K52)
KFCA. Helicobacter pylori-induced duodenuitis
KFCB. Chemical duodenitis
0. Alcoholic duodenitis (K29.1)
Inflammation of the duodenal mucosa due to alcoholic use

1. Drugs and medicaments
KFCC. Other specific causes of duodenitis
Eosinophilic/allergic
Lymphocytic
KFCD. Infectious duodenitis coded elsewhere
Bacterial
(except. Helicobacter pylori-induced)
(incl. duodenal phlegmone, dodenal abscess)
Viral
Fungal
Parasitic
KFCE. Due to systemic disorders classified elsewhere
0. Sarcoidosis
1. Vasculitis
(excludes: Crohn's disease K52)
KFCF. Due to other external causes
(Use external cause code *(Chapter XX), if desired,
to identify cause.)
0. radiation/thermal
1. foreign body
2. due to endoscopic procedures
etc
KFCG. Duodenitis, etiology unspecified, with specific endoscopic or pathologic features
0. Acute haemorrhagic duodenitis
Acute (erosive) duodenitis with hemorrhage
1. Granulomatous gastritis (NOS)
KFCH. Other and unspecified duodenitis

KFD Duodenal ulcer and erosion
(includes: Peptic ulcer disease in the duodenum, mucosal defect, ulcer scar,
Zollinger-Ellison syndrome, gastric erosions)
(excludes: Dieulafoy ulcer K29.52)
KFDA. Duodenal erosion
KFDB. Duodenal ulcer Helicobacter pylori-induced
KFDC. Duodenal ulcer drug/toxic-related
(includes NSAIDs ulcer)
(Use additional code if desired, to identify cause)
KFDD. Duodenal ulcer due to both Helicobacter pylori and drugs
KFDE. Other specific causes of duodenal ulcer
(excludes: Crohn's disease)
0. Zollinger-Ellison syndrome

1. Eosinophilic
2. Lymphocytic
3. Infectious diseases coded elsewhere
Bacterial
Viral
Fungal
Parasitic
4. Related systemic diseases
Sarcoidosis
Vasculitis
(excludes: Crohn's disease K52)
5. Stress ulcer and ulcerations (intensive care related)
6. Other external causes
Iatrogenic
(Use external cause code *(Chapter XX), if desired,
to identify cause)

KFDF. Duodenal ulcer, etiology unspecified, without complication (idiopathic)
0. Acute duodenal ulcer
(includes. Duodenal ulcer: acute without haemorrhage or perforation K26.3)
1. Chronic gastric ulcer
(includes. Duodenal ulcer: chronic without haemorrhage or perforation K26.7)
2. Dodenal ulcer: unspecified as acute or chronic, without haemorrhge or perforation K26.9)
KFDG. Duodenal ulcer etiology unspecified, with complication
0. with hemorrhage
00. acute with haemorrhage (K26.0)
01. chronic or unspecified with haemorrhage (K26.4)
1. with anemia (with occult bleeding)
2. with perforation
20. acute with perforation (K26.1)
21. acute with both perforation and haemorrhage (K26.2)
22. chronic or unspecified with perforation (K26.5)
23. chronic or unspecified with both perforation and haemorrhage (K26.6)
3. with obstruction or stenosis
KFDH. Duodenal ulcer etiology unspecified, located at
(location code could be used in combination with other codes in KFD as double code, if desired)
0. Bulbar portion