Directory of Life-Limiting conditions

Hain and Devins, Cardiff, 2011

The Directory of Life-Limiting Conditions is a list of nearly four hundred ICD10 codes associated with diseases that can limit life in children. The classification of a condition as 'life-limiting' means that its trajectory can plausibly be described by at least one of the archetypes set out in the 1997 ACT/RCPCH guidelines (1). The list for the Directory was obtained in 2011 by pooling diagnostic data from a number of children's hospices and specialist palliative medicine teams across the UK and combining it with data from death certificates (2).

It is important to remember that:

- **The Directory is not exhaustive**. There are conditions that would fulfil the criterion for 'life-limiting' that are not on the list simply because they were too rare to be captured by this method.
-
- **The Directory is not determinative.** It is a list of conditions that can limit life, not a list of children who should be referred to specialist palliative care. Not every child with a condition on this list will need specialist palliative care at any one time and indeed some will never need it. This is because a) some of the conditions can present with a range of severity, such as cerebral palsy b) Even children who will need palliative care at some point might not need it yet c) not all palliative care needs to be provided by specialist teams.

- **The Directory is not definitive.** What is considered 'life-limiting' involves a degree of subjective judgement which is influenced by current understandings and by the availability of technology, both of which can change with time. The advent of gene therapy, for example, might make many conditions in the Directory curable and no longer appropriate for inclusion. Acute trauma such as road traffic accidents, on the other hand, are not currently considered to be 'life-limiting conditions' but there are cogent reasons to consider that they should be in the future.

For any enquiries about the Directory (including comments or additions) please contact Dr Richard Hain richard.hain@southwales.ac.uk

1. ACT/RCPCH. A guide to the development of children's palliative care services. 1 ed. Bristol and London: ACT/RCPCH; 1997.
2. Hain et al.: Paediatric palliative care: development and pilot study of a 'Directory' of life-limiting conditions. BMC Palliative Care 2013 12:43. http://www.biomedcentral.com/1472-684X/12/43

Dictionary of Life Limiting Conditions (Hain, Devins et al 2011)

ICD 10 Code	ICD Name	Diagnosis
A		
A17	Tuberculosis of nervous system	Cerebral Tuberculosis
A81.0	Creutzfeldt-Jakob disease	Creutzfeld Jacob Disease
A81.1	Subacute sclerosing panencephalitis	SSPE
B		
B20-B24	Human immunodeficiency virus [HIV] disease	HIV
C		
C00-C97	Malignant neoplasms	
D		
D33	Benign neoplasm of brain and other parts of central nervous system	Brain tumour
D43	Neoplasm of uncertain or unknown behaviour of brain and central nervous system	Neoplasm of uncertain or unknown behavior, Brain supratent
D44.4	Neoplasm of uncertain or unknown behaviour of Craniopharyngeal duct	Craniopharygioma
D48	Neoplasm of uncertain or unknown behavior, bone and articular cartilage	
D56.1	Beta thalassaemia	Thalasaemia Major
D61.0	Constitutional aplastic anaemia	Aplastic anaemia
D61.9	Aplastic anaemia, unspecified	Medullary hypoplasia
D70	Agranulocytosis	Severe chronic neutropenia
D76.1	Haemophagocytic lymphocystosis	
D81	Combined immunodeficiencies	Immune Deficiency Syndrome
D82.1	Di George's syndrome	Di George syndrome
D83	Common variable immunodeficiency	Common variable immunodeficiency

D89.1	Cryoglobulinaemia	Cryoglobulinaemia
E		
E31.0	Autoimmune polyglandular failure	Autoimmune polyglandular failure
E34.8	Other specified endocrine disorders	Progeria
E70.2	Disorders of tyrosine metabolism	Tyrosinaemia
E71	Disorders of branched-chain amino-acid metabolism and fatty-acid metabolism	Maple syrup urine disease Hyperleucine-isoleucineaemia Methylmalonic acidaemia Proprionic acidaemia Adrenoleukodytrophy(Addison-schilder) Muscle carnitine palmityltransferase deficiency
E72	Other disorders of amino-acid metabolism	Fanconis syndrome Cerebral ocular renal disorder Cystinosis Lowes syndrome Molybdenum cofactor deficiency Sulphite oxidase deficiency Arginosuccinic aciduria Citrullinaemia Glutaric aciduria Methyl malonic aciduria Glycine encephalopathy Non ketotic hyperglycinamia Argininaemia Aminoaciduria
E74	Other disorders of carbohydrate metabolism	Glycogen storage disease type 1b Pompes disease

		Galactosaemia
		Pyruvate Dehydrogenase Deficiency
		Oxalosis renal failure
E75	Disorders of sphingolipid metabolism and other lipid storage disorders	Gangliosidosis
		Sandhoffs Disease
		GM2 Gangliosidosis (Tay Sachs)
		Tay Sachs Disease, infantile
		GM1 Gangliosidosis,early infantile
		GM1 Gangliosidosis, late infantile
		Other gangliosidosis
		Gangliosisdosis NOS
		Gangliosidosis GM1
		Gangliosidosis GM3
		Mucolipidosis 4
		Canavans leucodystrophy
		Gauchers Disease Type 2
		Metachromic Leukodystrophy juvenile
		Niemann Pick type a
		Niemann Pick type b
		Niemann Pick type c
		Other sphingolipidosis
		Fabry(Anderson) disease
		Gaucher Disease
		Krabbe Disease
		Niemann Pick Disease
		Farbers Disease

		Metachromic leukodystrophy
		Pelizaeus Merzbacher
		Sulfatase deficiency
		Batten Disease infantile
		Batten Disease late infantile
		Neuronal ceroid lipofucinosis
		Batten disease
		Bielschowsky-Jansky Disease
		Kufs disease
		Spielmeyer Vogt Disease
		Other lipid storage disorder
		Cerebrotendious cholesterosis (van Bogaert-Scherer-Epstein)
		Wolmans Disease
E76	Disorders of glycosaminoglycan metabolism	MPS1 Hurler
		MPS2 Hunter
		Other MPS
		Beta glucuronidase deficiency
		MPS types 3,4,6,7
		Marotaeux Lamy syndrome
		Moriquio syndrome
		Sanfillipo syndrome
E77	Disorders of glycoprotein metabolism	Pseudo hurlers/mucoplipdosis type 3
		Defects in post translational modification of lysosomal enzym
		Mucolipdosis 2 (I cell)
		Mucolipidosis 3
		Fucidosis

		CDG/CTG syndrome
		I cell disease
E79.1	Lesch-Nyhan syndrome	Lesch Nyhan
E83.0	Disorders of copper metabolism	Menkes
E84	Cystic fibrosis	Cystic Fibrosis
E88.0	Disorders of plasma-protein metabolism, not elsewhere classified	Alpha 1 antitrypsin deficiency
		Bisalbuninaemia
E88.1	Lipodystrophy, not elsewhere classified	Bernardnelli Lipodstrophy
F		
F80.3	Acquired aphasia with epilepsy [Landau-Kleffner]	Landau Kleffner syndrome
F84.2	Rett's syndrome	Retts syndrome
G		
G10	Huntington's disease	Huntingtons chorea, juvenile
G11.1	Early-onset cerebellar ataxia	Spinocerebellar ataxia
		Friedrichs ataxia
G11.3	Cerebellar ataxia with defective DNA repair	Ataxia telangectasia
G12	Spinal muscular atrophy and related syndromes	SMA type 1/Werdnig hoffman
		Chronic SMA
		Faziolondes syndrome
G20	Parkinson's disease	Parkinsons disease
G23.0	Hallervorden-Spatz disease	Hallervorden Spatz
G23.8	Other specified degenerative diseases of basal ganglia	Olivopontocerebellar Atrophy
G31.8	Other specified degenerative diseases of nervous system	Alpers syndrome
		Leigh Syndrome
G31.9	Degenerative disease of the nervous system unspecified	Aicardia-Goutieres
		Worster drought syndrome

G35	Multiple sclerosis	MS
G40.3	Generalized idiopathic epilepsy and epileptic syndromes	Familial myoclonic epilepsy
G40.4	Other generalized epilepsy and epileptic syndromes	Lennox gastaut West syndrome
G40.5	Special epileptic syndromes	Epilepsia partialis continuoa (Kozhevnikof)
G60.0	Hereditary motor and sensory neuropathy	Charcot-Marie-Tooth Hereditary motor sensory neuropathy
G60.1	Refsum's disease	Infantile refsum disease
G70.2	Congenital and developmental myasthenia	Congenital myasthaenia gravis
G70.9	Myoneuronal disorder, unspecified	
G71.0	Muscular dystrophy	DMD Gamma sarcoglycanopathy/limb girdle dystrophy Limb girdle dystrophy/type 2c sarcoglycaopathy
G71.1	Myotonic disorders	Myotonia chondrodystrophic Myotonia drug induced Myotonia symptomatic Myotonia congenital NOS Myotonia congenital dominant (thomsen) Myotonia congenital recessive (becker) Neuromyotonia (Isaacs) Paramyotonia congenital Pseudomyotonia Myopathycongenital
G71.2	Congenital myopathies	Congenital muscular dystrophy Congenital myopathies, disease central core Congenital myopathies,disease minicore Congenital myopathies, disease multicore

		Congenital myopathies, fibre type disproportion
		Myopathy:myotubular
		Myopathy:nemaline
G71.3	Mitochondrial myopathy, not elsewhere classified	Mitochondrial myopathy
G80.0	Spastic quadriplegic cerebral palsy	Spastic quadriplegic cerebral palsy
G80.8	Other cerebral palsy	Other CP
		Mixed CP syndromes
G82.3	Flaccid tetraplegia	
G82.4	Spastic tetraplegia	Spastic tetraplegia
G82.5	Tetraplegia, unspecified	Tetraplegia, unspecified
G93.4	Encephalopathy, unspecified	Recurrent encephalopathy
G93.6	Cerebral oedema	Cerebral oedema
G93.7	Reye's syndrome	Reye’s syndrome
H		
H11.1	Conjunctival degenerations and deposits	Multiple pterygium syndrome
H49.8	Other paralytic strabismus	Kearns Sayre syndrome
I		
I21	Acute myocardial infarction	Heart attack (9 days old)
I27.0	Primary pulmonary hypertension	Primary pulmonary HT
I42	Cardiomyopathy	Congestive cardiomyopathy
		Cardiomyopathy, hypertrophic
		Other hypertrophic cardiomyopathy
		Nonobstructive hypertrophic cardiomyopathy
		Endocardial fibroelastosis
		Congenital cardiomyopathy
		Other cardiomyopathies

		Restrictive cardiomyopathy
I61.3	Intracerebral haemorrhage in brain stem	4TH Ventricle Brain stem damage
I81	Portal vein thrombosis	Blocked portal vein
J		
J84.1	Other interstitial pulmonary diseases with fibrosis	Fibrosing alveolitis
J96	Respiratory failure, not elsewhere classified	Respiratory failure
J98.4	Other disorders of lung	Calcification of lung
		Cystic lung disease (acquired)
		Pulmolithiasis
K		
K55.0	Acute vascular disorders of intestine	Acute fulminate ischaemic colitis
		Acute intestinal infarction
		Acute small intestine ischaemia
		Mesenteric artery embolism
		Mesenteric artery infarction
		Mesenteric artery thrombosis
		Subacute ischaemic colitis
K55.9	Vascular disorder of intestine, unspecified	Ischaemic colitis NOS
K72	Hepatic failure, not elsewhere classified	Liver failure
K74	Fibrosis and cirrhosis of liver	Cirrhosis of liver
K76.5	Hepatic veno-occlusive disease	Venous occlusive disease
K86.8	Other specified diseases of pancreas	Schwachmann diamond
M		
M31.3	Wegener's granulomatosis	Wegeners granulomatosis
M32.1	Systemic lupus erythematosus with organ or system involvement	Lupus ? hypergammaglobulinaemia
M89.5	Osteolysis	Gorhams syndrome

N		
N04	Nephrotic syndrome	Congenital nephrotic syndrome
N17	Acute renal failure	Renal failure end stage
N18	Chronic renal failure	CRF
N19	Unspecified renal failure	Unspecified renal failure
N25.8	Other disorders resulting from impaired renal tubular function	ARC syndrome
P		
P10.1	Cerebral haemorrhage due to birth injury	Cerebral haemorrhage due to birth injury
P11.2	Unspecified brain damage due to birth injury	Unspecified brain damage due to birth injury
P20.0	Intrauterine hypoxia first noted before onset of labour	
P20.1	Intrauterine hypoxia noted during labour and delivery	
P21.0	Severe birth asphyxia	Severe birth asphyxia
P21.9	Birth asphyxia unspecified	
P28.5	Respiratory failure of newborn	
P29.0	Neonatal cardiac failure	
P29.3	Persistent fetal circulation	
P35.0	Congenital rubella syndrome	Congenital Rubella syndrome
P35.1	Congenital cytomegalovirus infection	Congenital CMV
P35.8	Other congenital viral diseases	Congenital varicella
P37.1	Congenital toxoplasmosis	Congenital toxoplasmosis
P52.4	Intracerebral haemorrhage of the fetus and newborn	
P52.5	Subarachnoid haemorrhage of the fetus and newborn	
P52.9	Intracranial haemorrhage of the fetus and newborn, unspecified	
P83.2	Hydrops fetalis not due to haemolytic disease	
P91.2	Hydrops fetalis NOS	
P91.6	Hypoxic ischaemic encephalopathy of newborn	Hypoxic brain damage
P96.0	Congenital renal failure	Congenital renal failure

Q

Q00.0	Anencephaly	Anencephaly
Q01	Encephalocele	Encephalocele Meningocele
Q03.1	Atresia of foramina of Magendie and Luschka	Dandy walker syndrome
Q03.9	Congenital hydrocephalus, unspecified	
Q04.0	Congenital malformations of corpus callosum	Acro colossal syndrome Aicardi syndrome
Q04.2	Holoprosencephaly	Holosprosencephaly
Q04.3	Other reduction deformities of brain	Hydranencephaly Lissencephaly Microgyria Pachgyria Agenesis of part of the brain Aplasia of part of the brain Hypoplasia of part of the brain Agyria
Q04.4	Septo-optic dysplasia	Septooptic dysplasia
Q04.6	Congenital cerebral cysts	Brain cyst Schizzencephaly
Q04.9	Congenital malformation of brain unspecified	Congenital anomaly NOS of brain Congenital deformity NOS of brain Congenital disease or lesion of brain NOS Congenital multiple anomalies NOS of brain
Q07.0	Arnold-Chiari syndrome	Arnold chiari malformation type1 Arnold chiari malformation type 2 Arnold chiari malformation type 3

		Arnold chiari malformation type 4
		Common arterial trunk
Q20.0	Common arterial trunk	Persistent truncus arteriosus
		Discordant ventriculoarterial connection
Q20.3	Discordant ventriculoarterial connection	Dextrotransposition of aorta
		Transposition of great vessels
		Double inlet ventricle
Q20.4	Double inlet ventricle	Common ventricle
		Cor triloculare biatriatum
		Single ventricle
Q20.6	Isomerism of atrial appendages	Isomerism of atrial appendages with asplenia or polysplenia
Q20.8	Other congenital malformations of cardiac chambers and connections	AS small left ventricle
Q21.3	Tetralogy of Fallot	Tetraology of fallot
Q21.8	Other congenital malformations of cardiac septa	Eisenmeyers defect
		Pentalogy of fallot
Q22.0	Pulmonary valve atresia	Congenital Pulmonary atresia
Q22.1	Congenital pulmonary valve stenosis	Pulm stenosis small vent
Q22.4	Congenital tricuspid stenosis	Tricuspid atresia
Q22.5	Ebstein's anomaly	Ebsteins anomaly
Q22.6	Hypoplastic right heart syndrome	Hypoplastic left heart syndrome
Q23.0	Congenital stenosis of aortic valve	Congenital stenosis of aortic valve
		Congenital aortic atresia
		Congenital aortic stenosis
Q23.2	Congenital mitral stenosis	Congenital mitral stenosis
		Congenital mitral atresia
Q23.4	Hypoplastic left heart syndrome	Hypoplastic left heart syndrome

		Atresia or marked hypoplasia of aortic valve with hypoplasia c atresia)
Q23.9	Congenital malformation of aortic and mitral valves, unspecified	Congenital malformation of aortic and mitral valves unspecifie
Q25.4	Other congenital malformations of aorta	Absence of aorta
		Aplasia of aorta
		Congenital aneurysm of aorta
		Congenital dilation of aorta
		Aneurysm of sinus of valsalva (ruptured)
		Double aortic arch(vascular ring)
		Hypoplasia of aorta
		Persistent convulsions of aortic arch
		Persistent right aortic arch
Q25.6	Stenosis of pulmonary artery	Stenosis of pulm artery
Q26.2	Total anomalous pulmonary venous connection	Total anamolous pulmonary venous connection
Q26.4	Anomalous pulmonary venous connection, unspecified	Anomalous pulm venous connection, unspecified
Q26.8	Other congenital malformations of great veins	Other congenital malformations of great veins
		Absence of vena cava
		Azygous continuoation of inferior vena cava
		Persistent left posterior cardinal vein
		Scimitar syndrome
Q28.2	Arteriovenous malformation of cerebral vessels	Bilateral AV malformation
Q32.1	Other congenital malformations of trachea	Atresia of trachea
		Congenital stenosis of trachea
Q33.6	Hypoplasia and dysplasia of lung	Hypoplasia and dysplasia of lung
Q39.8	Other congenital malformations of oesophagus	Absent oesphagus
Q41.0	Congenital absence, atresia and stenosis of duodenum	Dudodenal atresia
Q41.9	Congenital absence, atresia and stenosis of small intestine, part unspecified	Congenital absence, atresia and stenosis of intestine, part uns

		Congenital absence, atresia and stenosis of intestine NOS
Q43.1	Congenital (aganglionic) megacolon	Total intestinal aganglio
Q43.7	Persistent cloaca	Cloacal abnor exopthalmos
Q44.2	Atresia of bile ducts	Congenital biliary atresia
Q44.5	Other congenital malformations of bile ducts	Intrahepatic biliary hypoplasia
Q44.7	Other congenital malformations of liver	Alagilles biliary atresia
Q60.1	Renal agenesis, bilateral	Renal Failure congenital abscence of kidneys
Q60.6	Potter's syndrome	Potters syndrome RF
Q61.4	Renal dysplasia	Renal dysplasia
Q61.9	Cystic kidney disease, unspecified	Cystic kidney disease, unspecified
		Meckel-Gruber syndrome
Q64.2	Congenital posterior urethral valves	RF-urethral valves
Q74.3	Arthrogryposis multiplex congenita	Penn Shokeir syndrome
Q74.8	Other specified congenital malformations of limb(s)	Larsens syndrome
Q75.0	Craniosynostosis	Pfeiffer syndrome
Q77.2	Short rib syndrome	Short rib syndrome
		Asphyxiating thoracic dysplasia(Jeune)
Q77.3	Chondrodysplasia punctata	Conradis syndrome
Q77.4	Achondroplasia	Achondroplasia severe resp prob
Q78.0	Osteogenesis imperfecta	Osteogenesis Imperfecta
Q78.5	Metaphyseal dysplasia	Spendylo metaphyseal dysplasia
Q79.2	Exomphalos	Exopthalmos
		Omphalocele
Q79.3	Gastroschisis	Gastroschisis
Q80.4	Harlequin fetus	Harloquinn icthyosis
Q81	Epidermolysis bullosa	Dystrophic epidermolysis bullosa
		epidermolysis bullosa

		epidermolysis bullosa, junctional
		epidermolysis bullosa, herlitzs junctional
		Johanna Blizzard Syndrome
Q82.1	Xeroderma pigmentosum	Xeroderma pigmentosum
Q82.4	Ectodermal dysplasia (anhidrotic)	LADD-EEC ectodermal dysplasia
Q85.1	Tuberous sclerosis	Tubero sclerosis
Q85.8	Other phakomatoses, not elsewhere classified	Sturge Weber syndrome
Q86.0	Fetal alcohol syndrome (dysmorphic)	Foetal alcohol syndrome
Q87.0	Congenital malformation syndromes predominantly affecting facial appearance	Pierre robin syndrome
		Acrocephalopolysyndactly
		Acrocephaolsyndactly(Apert)
		Cryptophtalmos syndrome
		Cyclopia
		Goldenhar syndrome
		Moebius syndrome
		Oro-facial-digital syndrome
		Robin syndrome
		Whistling face syndrome
Q87.1	Congenital malformation syndromes predominantly associated with short stature	Cornelia de lange
		Aarskog syndrome
		Cockayne syndrome
		Dubowitz syndrome
		Robinhow-silverman-smith syndrome
		Russel-silver syndrome
		Seckel syndrome
		Smith-lemli-opitz syndrome

		Pradar Willi Syndrome
Q87.2	Congenital malformation syndromes predominantly involving limbs	Rubenstein Tabyii
Q87.8	Other specified congenital malformation syndromes, not elsewhere classified	Alport syndrome
		Laurence Moon (Bardet)Biedl syndrome
		Zellweger syndrome
Q91	Edwards' syndrome and Patau's syndrome	Edwards syndrome trisomy 18
		Trisomy 13 Patau
Q92.0	Whole chromosome trisomy, meiotic nondisjunction	Trisomy 22
		Trisomy 9
Q92.1	Whole chromosome trisomy, mosaicism (mitotic nondisjunction)	Trisomy 10 with deletion
		Chr 8 duplication
Q92.4	Duplications seen only at prometaphase	Chromosomal abnormality duplication of X
Q92.7	Triploidy and polyploidy	Tetrasomy 5p mosaicism
Q92.8	Other specified trisomies and partial trisomies of autosomes	
Q93.2	Chromosome replaced with ring or dicentric	Inversion chr 10
Q93.3	Deletion of short arm of chromosome 4	Deletion of short arm chrom 4
		Wolff Hirschorn syndrome
Q93.4	Deletion of short arm of chrom 5	Cri du chat
		Jacobsen syndrome (11q deletion)
Q93.5	Absecence long arm chr 14	9p minus syndrome
		Angelamn's Syndrome
		Williams syndrome
		Chromoseme deletion2
Q93.8	Other deletions from the autosomes	DeGouchy syndrome
Q95.2	Balanced autosomal rearrangement in abnormal individual	Translocation 14/22

Q99.2	Fragile X chromosome	Fragile X PVS
R		
R06.8	Other and unspecified abnormalities of breathing	Ondines curse
T		
T86.0	Bone-marrow transplant rejection	Chronic GVHD
T86.2	Heart transplant failure and rejection	Failed Heart transplant
Z		
Z51.5	Palliative care	Palliative Care