ANZICS

ANZICS Centre for Outcome and Resource Evaluation

ANZPIC Registry
Chronic Condition Definitions

AUSTRALIAN AND NEW ZEALAND PAEDIATRIC INTENSIVE CARE REGISTRY

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Complex chronic conditions include any medical condition that can be reasonably expected to last at least 12 months (unless death intervenes) and to involve either several different organ systems or one organ system severely enough to require specialty paediatric care and probably some period of hospitalisation in a tertiary care centre. The condition must have been diagnosed prior to ICU admission.

Category	Subcategory	Examples, further notes
Congenital Anomalies		
Congenital malformations and deformations of the musculoskeletal system	Congenital malformations of spine and bony thorax	Scoliosis, Spina bifida occulta, Klippel-Feil syndrome, Congenital spondylolisthesis, Cervical rib, Osteopetrosis, Diaphyseal dysplasis, Jeune syndrome (Asphyxiating thoracic dystrophy)
	Congenital malformation of skull and face bones	Craniosynostosis, Hypertelorism, Pierre-Robin Syndrome, Crouzons Syndrome/Craniofacial Dysostosis, Aperts, Pfeiffer, Saethre-Chotzen
	Osteodystrophies and osteogenesis imperfecta	Osteochrondrodysplasia, Osteopetrosis, Albers-Schonberg disease, Enchondromatosis, Polyostotic fibrous dysplasia, Diaphyseal dysplasia
Chromosomal abnormalities	Sex chromosome anomalies	Gonadal Dysgensis, Turner Syndrome, Kleinfelter Syndrome, Fragile X Syndrome
	Autosomal monosomies, deletions/translocations/anomalies	Cri-du-chat syndrome
	Trisomies and partial trisomies of autosomes	Trisomy 13, 18 or 21
	Congenital malformation syndromes involving early overgrowth	Beckwith-Wiedermann, Sotos
Phakomatoses	Neurocutaneous	Neurofibromatosis, Tuberous sclerosis, Peutz-Jegher, Sturge-Weber, Von Hippel Lindau, Hamartoses (except lymphoedema or vascular)
Other congenital malformation syndromes	Congenital malformation syndromes affecting multiple systems	Marfan's Syndrome, Alport's Syndrome, McCune-Albright syndrome
	Congenital malformation syndromes predominantly associated with small stature	Noonan's Syndrome
Diaphragm and abdominal wall	Congenital malformations of diaphragm	Diaphragmatic hernia
	Congenital malformations of the abdominal wall	Exomphalos, Gastroschisis, Omphalocele, Prune belly syndrome/Eagle-barrett syndrome
Connective tissues	Ehlers Danlos syndrome	

Category	Subcategory	Examples, further notes
Congenital exposure syndromes	Syndromes due to in-utero exposure to drug or toxin	Foetal alcohol syndrome, foetal phenytoin syndrome
Other congenital anomalies	Teratology	Conjoined twins
	Ectodermal dysplasia	
	Connective tissue	Epidermolysis bullosa, Ehlers Danlos syndrome
	Multiple congenital malformations, not elsewhere classified	VACTERL association
Premature/Neonatal *(Note: Applies only to patients aged up to 1 year (chronological age) at time of ICU admission)*		
Disorders relating to length of gestation and foetal growth	Newborn light or small for gestational age < 750g	
	Gestation age <= 26 completed weeks only	
Cerebral injury and other cerebral disturbances during the perinatal period	Cerebral haemorrhage, subdural haemorrhage or tentorial tear due to birth trauma	
	Unilateral IVF >= grade 3	
	Birth injury to spine or spinal cord	
	Hypoxic ischaemia encephalopathy (HIE) - any	
	Kernicterus due to isoimmunization or other specified reason	
	Neonatal cerebral leucomalacia	
Respiratory disorders specific to the perinatal period	Chronic respiratory disease originating in newborn period (excluding chronic lung disease as a consequence of mechanical respiration)	Bronchopulmonary Dysplasia (BPD), Wilson-Mikity Syndrome
Congenital infections	Congenital viral disease	Congenital rubella, CMV, Herpes simplex, Varicella, HIV, Hepatitis B
	Other congenital infections	Toxoplasmosis, syphilis
Other disorders originating in the perinatal period	Necrotising enterocolitis stage 3	Stage 3: Advanced NEC: severely ill, marked distension, signs of peritonitis, hypotension, metabolic & respiratory acidosis, DIC, +/- bowel perforation.
	Hydrops Foetalis due to haemolytic disease or other	
Malignancy *(Note: All malignancies are considered chronic until 5 years post last treatment)*		
Malignancy	Malignant neoplasms - all sites	Ewings, Neuroblastoma, Phaemochromocytoma, Lymphoma and Leukaemia
	Neoplasms of uncertain or unspecified behaviour, any site	
	Neurofibromatosis (non-malignant)	
	Chemotherapy: via any route	
Cardiovascular		
Heart	Absent pulmonary valve	

Category	Subcategory	Examples, further notes
	Aortic valve or supravalvular atresia or stenosis	
	Aortic valve incompetence	
	AV canal defect – complete or partial including Primum ASD	
	DILV	
	DORV	
	Ebstein's anomaly	
	Hypoplastic left heart syndrome	
	Hypoplastic right heart syndrome	
	Left ventricular outflow obstruction	
	Mitral valve stenosis	
	Mitral valve incompetence	
	Pulmonary valve atresia or stenosis	
	Pulmonary valve incompetence	
	Tetralogy of Fallot – with or without RV outflow obstruction	
	Tricuspid valve atresia or stenosis	
	Tricuspid valve incompetence	
	Unrepaired ASD	Considered chronic until repaired
	Unrepaired VSD	Considered chronic until repaired
Vessels	Anomalous coronary artery from pulmonary artery	
	Anomalous pulmonary venous drainage – total or partial	
	Aortic atresia	
	Aortopulmonary window	
	Coarctation of the aorta	
	Interrupted or Hypoplastic aortic arch	
	MAPCAS	
	Pulmonary artery atresia or stenosis	
	Pulmonary Arterio-Venous Malformation (AVM)	
	Transposition of the Great Arteries – d or l	
	Truncus arteriosus	
	Unrepaired PDA	Considered chronic until repaired
Cardiomyopathies	Cardiomyopathy – any	
	Hypertrophic Obstructive Cardiomyopathy (HOCM)	
Acquired Cardiovascular Diseases	Cardiac tumour	
	Endocarditis	
	Kawasaki's disease with coronary aneurysms	
	Myocardial ischaemia or infarction	
	Pericarditis or effusion - chronic	
	Pulmonary hypertension	
	Rheumatic heart disease	

Category	Subcategory	Examples, further notes
	Vascular thrombosis or occlusion – chronic or recurrent	
	Vasculitis	
	Systemic hypertension	
	Valvular stenosis or incompetence	
Rhythm Disorders – recurrent or chronic	Supraventricular tachyarrhythmias	
	Ventricular tachyarrhythmias	Catecholaminergic polymorphic ventricular tachycardia (CPVT), Brugada syndrome, Congenital long QT syndromes (Jervell and Lange-Nielson, Romano-Ward), Short QT syndrome
	Heart block – congenital or acquired	
	Sick sinus syndrome	
Haematologic/Immunologic		
Anaemias	Anaemia due to enzyme disorders	G6PD deficiency
	Thalassaemia	
	Sickle-cell disorders	
	Other aplastic anaemias and other bone marrow failure syndromes	
Immunodeficiency	Functional disorders of polymorphonuclear neutrophils	Chronic granulomatous disease
	Immunodeficiency with predominantly antibody defects	Selective deficiencies of immunoglobulins or hyper IgM etc.
	Combined immunodeficiencies	Severe combined immunodeficiency (SCID), Wiskott-Aldrich syndrome, Ataxia Telangiectasia, DiGeorge syndrome (22q11del)
	Common variable immunodeficiency	
	Human immunodeficiency virus [HIV] disease	
	Congenital agranulocytosis	Kostmann syndrome
	Cyclic neutropaenia	
	Other imunodeficiency	Complement defects
Coagulation/haemorrhagic	Hereditary factor VIII deficiency	Hemophilia A
	Hereditary deficiency of other clotting factors	Factor IX deficiency = Hemophilia B (aka Christmas disease), von Willebrand's disease, deficiencies of Factors I, II, V, VII, X, XIII.
	Congenital and hereditary thrombocytopaenia purpura	
Hemophagocytic Syndromes	Hemophagocytic lymphohistiocytosis	Familial or genetic HLH
	Haemophagocytic syndrome, infection-associated	
	Other histiocytosis syndromes	Langerhans's Cell Histiocytosis (LCH). Other synonyms for this include: Histiocytosis X, Eosinophilic granuloma, Letterer-Siwe disease, Hand-Schüller-Christian syndrome

Category	Subcategory	Examples, further notes
Diffuse diseases of connective tissue	Wegener's granulomatosis	
	Systemic lupus erythematosus	
	Mucocutaneous lymph node syndrome (Kawasaki)	
Other haematologic/ immunologic	Resection of spleen	
Respiratory		
Airway	Bronchogenic cyst	
	Bronchomalacia	
	Laryngeal cleft	
	Laryngeal web	
	Subglottic stenosis	
	Tracheal rings	
	Tracheal stenosis	
	Tracheal web	
	Tracheoesophageal fistula	
	Tracheomalacia	
Lungs	Alveolocapillary dysplasia	
	Congenital diaphragmatic hernia	
	Congenital lobar emphysema	
	Cystic adenomatoid malformation	
	Cystic fibrosis	
	Lymphangiectasis	
	Pulmonary hypoplasia	
	Pulmonary sequestration (including Scimitar syndrome)	
Other respiratory	Alpha 1 antitrypsin deficiency	
	Central hypoventilation syndrome	Ondine’s curse
	Ciliary dyskinesia (including Kartagener syndrome)	
	Surfactant protein deficiency	
Chronic respiratory diseases	Asthma	Considered if one previous inpatient admission in the last 12 months with asthma OR ever been intubated with asthma
	Bronchiectasis/chronic suppurative lung disease	
	Chronic lung disease/ Bronchopulmonary dysplasia	
	Pulmonary fibrosis	
	Restrictive lung disease	
Respiratory surgery	Prior lobectomy	
Neurologic/Neuromuscular		
Brain and spinal malformations	Malformations of the brain	Anencephaly, Holoprosencephaly, Encephalocoele, Microcephaly, Hydrocephalus, Chiari malformation, Lissencephaly,

Category	Subcategory	Examples, further notes
	Malformations of the spinal cord	Syringomyelia, Spina bifida, Tethered cord
	Malformations of the nervous system	
	Familial dysautonomia	
CNS: Central storage disorders	Mucopolysaccharidoses	Hunter / Hurler / Scheie / Sanfillipo / Morquio / Maroteaux-Lamy / Sly / Hyaluronidase deficiency
	Sphingolipidoses	GM1 / Tay-Sachs / Fabry / Gaucher / Niemann-Pick / Krabbe / metachromatic lecodystrophy
	Other central storage disease	
CNS: Ataxias	Hereditary ataxia	
	Cerebellar ataxia	
CNS: Other	Any other neurodegenerative disease	Leigh syndrome
	Spinal muscular atrophy	
	Tuberous sclerosis	
	Rett syndrome	
	Malignant neuroleptic syndrome	
	Global developmental delay	
	Cerebral palsy	All types
	Intellectual disability	
Epilepsy	Any form of known epilepsy with status epilepticus	
	Any form of known epilepsy that is intractable	Intractable if failure of adequate trials of two tolerated and appropriately chosen and used AED schedules (whether as monotherapies or in combination) to achieve sustained seizure freedom (Kwan, Epilepsia 2009;51:1069–1077). Also: Dravet syndrome, Lennox Gastaut syndrome, West syndrome, Otahara syndrome
CNS: Demyelination	Central pontine myelinolysis	
	Transverse myelitis	
	Other demyelinating disease of CNS	Multiple sclerosis, Devic's disease, ADEM, Acute haemorrhagic leucoencephalitis
CNS: Paralysis	Hemiplegia	
	Paraplegia	
	Quadriplegia	
	Locked in Syndrome	
	Paralytic syndromes	
	Hypoxic brain injury	
	Brain compression	
	Persistent vegetative state	
CNS: CSF shunt	Ventricle to atrium, pleura, peritoneum	Any CSF shunt, regardless of type or site
	Spinal canal to pleura, peritoneum	

Category	Subcategory	Examples, further notes
CNS: Surgery	Hemispherectomy	Any type, any approach
Occlusion of cerebral vessels	Cerebral venous thrombosis	
	Cerebral infarction	
	Cerebral haemorrhage	
Muscular dystrophies/myopathies	Any primary muscular dystrophy	Classical Duchenne, Becker, Fascioscapulohumeral, Limb-girdle and Myotonic
	Any myotonic disorder	
	Any primary disorder of muscle	Congenital myopathies including mitochondrial
	Myasthenia gravis	
Movement disorders	Any dystonia	
	Any chronic ataxia	Cerebellar ataxia, Hereditary ataxia, Congenital ataxia
	Any parkinsonian syndrome	
	Any extrapyramidal or movement disorder	Sydenham's chorea
	Huntington's chorea	
	SMA all types	
	Myoclonus	Any type
	Others	Restless legs syndrome, Stiff man syndrome, Any extrapyramidal movement disorder
	Hallevorden-Spatz syndrome	Any other basal ganglia degenerative disease
Gastrointestinal		
Congenital anomalies	Oesophagus	Oesophageal atresia with or without tracheo oesophageal fistula, Tracheo oesophageal fistula without atresia (H-type), Congenital stenosis or stricture, web
	Small intestine: congenital absence, atresia, stenosis	Duodenal, Ileal, Jejunal atresia/stenosis
	Large intestine: congenital absence, atresia, stenosis	Anal atresia/stenosis with and without fistula
	Other congenital malformations of intestine	Hirschsprung's disease, malrotation, intestinal duplication
	Congenital malformations of gallbladder, bile ducts and liver	Biliary atresia, Choledochal cyst
	Other congenital malformations of digestive system	Agenesis/aplasia/hypoplasia of pancreas
Chronic liver disease and cirrhosis		Chronic hepatitis, Cirrhosis and fibrosis, Autoimmune hepatitis, Central haemorrhagic necrosis of liver, Liver infarction, Hepatic veno occlusive disease
Other liver		Fatty liver, Chronic passive congestion of liver, Hepatopulmonary syndrome
Portal hypertension	With/without varices	
	Budd-Chiari syndrome	

Category	Subcategory	Examples, further notes
Inflammatory bowel diseases		Crohn's disease and ulcerative colitis
Chronic vascular disorders of intestine		Chronic ischemic entero/colitis; Chronic mesenteric ischemia; Chronic vascular insufficiency of intestine,
Volvulus		
Megacolon, not elsewhere classified		Megacolon, toxic megacolon
Acquired absence of stomach [or part of], or other parts of digestive tract	Small intestine or large intestine	Short gut syndrome, colectomy, hemicolectomy, ileal resection
Surgical resections		Tongue, Oesophagus, Stomach, Small intestine, Duodenum, Large intestine, Liver, Pancreas, NEC requiring surgical intervention
Dilatations and gastrointestinal intraluminal devices		Into oesophagus, Small intestine, Large intestine (stents)
Gastrostomy, Ileostomy, Colostomy & other artificial openings	Includes complications of these	
Oesophageal bypass procedures		Oesophagus to cutaneous, Oesophagus to stomach, intestine
Stomach bypass procedure		Stomach to cutaneous, stomach to intestine
Bypass other bowel		Ileum to cutaneous, Caecum to cutaneous, Colon to cutaneous
Renal/Urologic		
Congenital	Congenital malformations of the kidney or urinary system	Renal dysplasia, Renal agenesis, Any cystic Kidney disease, Other renal/urinary malformations, Congenital obstruction of renal pelvis, Posterior urethral valves
Chronic	Chronic renal failure/chronic kidney disease	
Bladder	Neuromuscular functional bladder problem	Neurogenic bladder, Neuropathic bladder
Acquired	Acquired absence of kidney	Traumatic or surgical nephrectomy
	Acquired absence of other urinary tract	Traumatic loss, surgical removal or bypass of other part of urinary tract
	Artificial urinary tract opening	Nephrostomy, Vesicostomy, Cystostomy, Appendicovesicostomy
Metabolic		
Amino Acid Metabolism	Classical phenylketonuria	PKU
	Other disorders of aromatic amino-acid metabolism	Disorders of phenylalanine or tyrosine but not PKU: Alkaptonuria, Tyrosinaemia, Albinism
	Maple-syrup-urine disease	
	Disorder of branched-chain amino-acid metabolism, unspecified	Methylmalonic acidaemia, Propionic acidaemia, Isovaleric acidaemia

Category	Subcategory	Examples, further notes
	Disorders of fatty-acid metabolism	Medium-chain acyl-coenzyme A dehydrogenase (MCAD) deficiency, Long-chain 3-hydroxyacyl-coenzyme A dehydrogenase (LCHAD) deficiency, Very long-chain acyl-coenzyme A dehydrogenase (VLCAD) deficiency
	Disorders of carnitine metabolism	Carnitine transport protein
	Peroxisomal disorders	Adrenoleukodystrophy, Refsum disease
	Disorders of amino-acid transport	Cysteinuria
	Disorders of urea cycle metabolism	OTC deficiency (orthinine transcarbamylase deficiency), Citrullinaemia, Argininosuccinic aciduria, Carbamoyl phosphate synthetase deficiency
	Disorder of amino-acid metabolism, unspecified	
Carbohydrate Metabolism	Glycogen storage disease	Pompe's disease, von Gierke's disease, McArdle disease
	Disorders of fructose metabolism	
	Disorders of galactose metabolism	Galactosemia (there are several enzymes that may be deficient)
	Disorders of pyruvate metabolism and gluconeogenesis	Pyruvate dehydrogenase deficiency, pyruvate carboxylase deficiency
	Other specified disorders of carbohydrate metabolism	
	Disorder of carbohydrate metabolism, unspecified	
Lipid Metabolism	Disorders of sphingolipid metabolism and other lipid storage disorders	GM1-gangliosidosis, GM2-gangliosidosis (infantile form =Tay-Sachs disease), Fabry disease, Gaucher disease, Metachromatic leukodystrophy, Krabbe disease, Niemann-Pick disease
	Hyperlipidaemia, unspecified	
	Lipodystrophy, not elsewhere classified	
Storage Disorder	Mucopolysaccharidosis, any type	Hurler syndrome, Hunter syndrome, Sanfilippo Morquio Maroteaux Lamy, Scheie, Sly, Hyaluronidase deficiency
Other Metabolic Disorders	Disorders of bilirubin excretion	Gilbert syndrome, Crigler-Najjar syndrome, Dubin Johnson syndrome
	Lesch-Nyhan syndrome	
	Other disorders of purine and pyrimidine metabolism	
	Other disorders of bilirubin metabolism	
	Disorder of bilirubin metabolism, unspecified	
	Disorders of copper metabolism	Menkes' syndrome, Wilson's disease
	Disorders of iron metabolism	

Category	Subcategory	Examples, further notes
	Multiple Carboxylase Deficiency, unspecified	Biotinidase deficiency
	Hypoglycaemia, unspecified	
	Disorders of mitochondrial oxidative phosphorylation, unspecified	MELAS (mitochondrial encephalopathy lactic acidosis and stroke-like episodes), Kearns-Sayre syndrome, Myoclonic epilepsy with ragged-red fibers (MERRF), Lebers optic neuropathy,
	Disorders of neurotransmitter metabolism, unspecified	
	Haemochromatosis, unspecified	
	Other and unspecified metabolic disorders	
Endocrine Disorders	Adrenocortical insufficiency, unspecified	
	Hypopituitarism	
	Diabetes insipidus	
	Syndrome of inappropriate secretion of antidiuretic hormone	
	Hypothalamic dysfunction, not elsewhere classified	
	Disorder of pituitary gland, unspecified	
	Drug-induced Cushing's syndrome	
	Cushing's syndrome, unspecified	
	Congenital adrenogenital disorders associated with enzyme deficiency	
	Adrenogenital disorder, unspecified	
	Congenial adrenal hyperplasia	
	Adrenal medullary dysfunction	
	Hypothyroidism	
	Thyroid disorder, unspecified	
	Diabetes mellitus, insulin dependent	
	Hypoparathyroidism	
Transplantation *(Notes: If transplant is regarded as curative, and ICU admission is post-transplant, then transplant only to be considered a chronic condition. If transplant does not cure the underlying disease (e.g. BMT for tyrosine kinase deficiency), then both transplant and underlying disease can be considered as chronic conditions.)*		
Transplantation - cardiac	Heart transplantation	
Transplantation - respiratory	Lung transplantation (lobe, single or double lung)	
Transplantation - renal	Kidney transplantation (related, unrelated)	
Transplantation - gastrointestinal	Liver transplantation (related, unrelated)	
	Intestinal transplantation (large, small)	

Category	Subcategory	Examples, further notes
	Pancreatic transplantation (non-autologous, autologous islet cell via any route)	
Transplantation - haematological	Marrow, Cord blood, Haemopoietic stem cells (autologous or non-autologous)	
Transplantation - miscellaneous	Transplant miscellaneous (e.g. splenic or other tissue/organ not listed)	
Technology Dependency *(Note: Must be considered chronic together with the system affected)*		
Neuro	CSF drainage device	
	Implanted neurostimulator lead	
	Synthetic substitute to ventricle or CSF drainage pathways (e.g., aqueductal stent placement)	
	Baclofen pump	
Cardiac	Prosthetic, bioprosthetic or biological heart valve	
	Conduit (prosthetic or biological)	
	Cardiac pacemaker (implantable) or contractility modulation device (delivers a biphasic signal to RV septum during absolute refractory period)	
	Automatic (implantable) cardiac defibrillator	
	Intravascular device including stents and implants	
	Heart assist device (BiVAD, LVAD, RVAD) or fully implantable artificial heart	
	Long-term vascular access device	
Respiratory	Tracheostomy	
	Tracheal/airway stent	
	Dependence on aspirator (vacuum) to remove airway secretions	
	Diaphragmatic pacemaker	
	Home CPAP or BIPAP	
	Home oxygen therapy	
	Home ventilation	
	Dependence on supplemental oxygen	
Renal	Dependence on renal dialysis (hemo or PD)	
	Cystostomy, vesicostomy, nephrostomy, ureterostomy, urethrostomy	
	Kidney pelvis or ureter to bladder bypass	

Category	Subcategory	Examples, further notes
	Arteriovenous fistula	
	Long-term vascular access device	
Gastro	Gastrostomy/PEG/other long-term feeding tube (e.g., NG or TPT)	
	Jejunostomy, Ileostomy, Colostomy, Any gut ostomy	
	Gastric lap band or similar	
	Oesophageal stent	
	Bypass oesophagus/upper oesophagus (e.g., gastric transposition or colon interposition for long seg oesophageal atresia)	
	Bypass stomach	
	Long-term vascular access device	
Metabolic	Pump external or internal for hormone infusion (e.g., insulin)	
Miscellaneous	Other internal orthopaedic devices, implants, and grafts	
	Spinal fusion of any type	
	External osteogenesis devices	Distraction osteogenesis (mandible, leg etc.)
	Dependence on supplemental oxygen	
Mental Health/Behavioural *(Notes: Include if the disorder is chronic (likely to last at least 12 months), and requires ongoing treatment (pharmacological/psychological/psychiatric))*		
Psychoactive substance use	Drug dependence (alcohol, amphetamines etc.)	
Schizophrenia, schizotypal, delusional, and other non-mood psychotic disorders	Schizophrenia (any form), or other non-mood psychotic disorder	
Mood (affective) disorders	Bipolar disorder	
	Depression	
Anxiety, dissociative, stress-related, somatoform and other nonpsychotic mental disorders	Anxiety or panic disorder	
	Post-traumatic stress disorder (PTSD)	
	Obsessive compulsive disorder (OCD)	
Behavioural syndromes associated with physiological disturbances and physical factors	Eating disorder (Anorexia nervosa, Bulimia)	
Disorders of personality and behaviour	Gender identity disorder	
Intellectual disabilities	Intellectual disability; any kind	
Pervasive and specific developmental disorders	Autism, Autism spectrum disorder, Asperger's syndrome	

Category	Subcategory	Examples, further notes
	Global developmental delay	
Behavioural and emotional disorders with onset usually occurring in childhood and adolescence	Attention deficit hyperactivity disorder (ADHD)	
	Oppositional defiant disorder (ODD)	
	Tourette's syndrome, Tic disorders	
	Behavioural and emotional disorder, other	