

# THE UNOFFICIAL GUIDE TO PAEDIATRICS

**FIRST EDITION**

**Zeshan Qureshi BM BSc (Hons) MSc MRCPCH**  
Academic Clinical Fellow, Great Ormond Street, UK and  
Institute of Global Health, UCL, UK  
Paediatric Registrar, London Deanery, UK



**ZESHAN  
QURESHI**

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# Introduction

It has been a privilege to work with so many Paediatricians, and to serve as part of the big teams that deliver the excellent care that every child deserves. Whilst my career to date has involved challenging situations, I have invariably been able to unite with colleagues and parents around the fact that above anything else, the wellbeing of a child should not be compromised.

It has been a privilege to work with so many Paediatricians, and to serve as part of the big teams that deliver the excellent care that every child deserves. Whilst my career to date has involved challenging situations, I have invariably been able to unite with colleagues and parents around the fact that above anything else, the wellbeing of a child should not be compromised.

Editing this book, and working closely with my professional colleagues has really made me reflect on what the true definition of a Paediatrician might be. I'm on a Paediatric training programme, but I don't think this is necessary to be a Paediatrician. I am privileged to say that I have passed the MRCPC membership examinations, but again don't think that is necessary to be a Paediatrician. I am now delighted to say that I've edited a Paediatrics textbook. But this doesn't qualify me as a Paediatrician.

So what is the core essence of this profession? Who can be a Paediatrician, in the true spirit of the word? And who should decide? In my humble opinion, it comes down to one simple litmus test. Can you do what is necessary, within the limitations of your knowledge, to be an advocate for a potentially sick child? Are you willing to try your utmost to communicate with a child and family to identify what their possible concerns are, and tease out any relevant pathology? If something goes wrong, or you are unhappy with something that is done regarding a child's care, regardless of any contextual factors, will you speak up on behalf of the child?

There is no substitute for clinical experience. Reading this book will inform you about Paediatrics. But to me, the most important thing in Paediatrics comes down to caring for the child, and when it comes down to this there should be no hierarchy: be tactful, use the appropriate channels, but never hesitate to speak up whenever you are worried that patient care is being compromised, regardless of who it might offend.

Anyone can be a Paediatrician. A medical student on a Paediatric rotation; the student will often take the opportunity

to spend more time listening to the patient than any healthcare professional that day: and I'm always grateful when a student comes up to me relaying valuable patient concerns and diagnostic information: they are a Paediatrician. The primary care physician that follows a child from womb to adulthood: they are a Paediatrician. Their knowledge of the family and the

**'...the most important thing in Paediatrics comes down to caring for the child, and when it comes down to this there should be no hierarchy'**

child throughout their life course is indispensable in identifying when things might go wrong in advance. The academics that improve the evidence on which care can be delivered: they are Paediatricians. The managers and policy makers that turn ideas into a reality: they are Paediatricians. And the Emergency Medicine doctor that sees a frightened parent and sick child for the first time, the ENT surgeon, the orthopaedic surgeon, the paediatric surgeon, the geneticist, the immunologist, the physiotherapist, the art therapist, the play specialist, the nurse, the dietician, the pharmacist, the social worker, the teacher, the police, every specialist, every person, every advocate that helps identify and address concerns and potential concerns to a child's wellbeing: They are all Paediatricians.

I am indebted to all their guidance and help in helping me provide care to children that I cannot fully provide on my own. It's up to you to decide what a Paediatrician is. But in my humble opinion, you can all be a Paediatrician today.

**Zeshan Qureshi**  
**Chief Editor**  
**Unofficial Guide to Paediatrics**

# The Unofficial Guide to Medicine Project

Additionally, we want you to get involved. This textbook has mainly been written by junior doctors and students just like you because we believe:

...that fresh graduates have a unique perspective on what works *for students*. We have tried to capture the insight of students and recent graduates to make the language we use to discuss this complex material more digestible for students.

...that texts are in *constant* need of being updated. *Every student* has the potential to contribute to the education of others by innovative ways of thinking and learning. This book is an open collaboration with you.

You have the power to *contribute* something valuable to medicine; we welcome your suggestions and would love for you to get in touch.

 Please get in touch and be part of the medical education project

 [admin@unofficialguidetomedicine.com](mailto:admin@unofficialguidetomedicine.com)

 [@DrZeshanQureshi](https://twitter.com/DrZeshanQureshi)

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# Foreword



## Dr Simon Broughton PhD FRCPCH

- Consultant Paediatrician, King's College Hospital NHS Foundation Trust
- Senior Lecturer, King's College London – Course Director, MSc in Advanced Paediatrics
- Training Programme Director

Congratulations to Zeshan and his colleagues on producing 'The Unofficial Guide to Paediatrics'. It is a huge piece of work by trainees and experts for anybody who has an interest in paediatrics, from medical students to established consultants and anybody interested in caring for children.

This book covers paediatrics in a traditional system based approach, but also has sections on the expanding speciality of adolescent healthcare, child health and the law, and public health. In addition, with sections on undergraduate and postgraduate assessments, starting out as a junior doctor and career sections, it provides useful advice to medical students and junior doctors wherever they are in their own career.

So, why do we need another textbook on paediatrics? There are already plenty of excellent texts on this subject, however none have created a book like this. The inspiration behind this book is the working together of junior doctors, medical students, and experts to pull together a textbook that is accessible to all types of learners. We now live in a world where knowledge is so widely and freely available, that simply reprinting knowledge is becoming unnecessary. If knowledge is to be pulled together in a textbook, then every effort should be made to make that knowledge as relevant and accessible to the reader as possible, and that is what the Unofficial Guide to Paediatrics achieves.

Every effort has been made to make this textbook as up to date as possible. However, inevitably, new research and guidance will be published. The genius behind this book however, is in empowering readers or users of this book to write to Zeshan with updates and suggestions for future editions.

Being a Paediatrician is an absolute privilege, caring for children and young people and their families at very difficult times in their lives is an unbelievably rewarding challenge. One of the challenges that busy paediatricians struggle with, is keeping themselves up to date in all areas of paediatrics. The Unofficial Guide to Paediatrics will help with that, providing guidance to paediatricians of the future and assist in providing excellent care to children, young people and their families.



## Beryl Lin

- President, University of New South Wales Medical Society
- Co-chair, University of New South Wales Paediatrics Special Interest Group

Paediatrics is a 'big' topic about 'little' people. It is intellectually challenging and an exciting field for research and learning, but it can also be daunting when medical school is often set up to focus on adult medicine. Caring for children is different – both in a purely scientific sense, but also the way a sick child and their family should be approached, the dynamic of the hospital and multidisciplinary team, the ethical and sociocultural considerations involved, and even the career pathways that present are unique – all of which are covered in this book.

The Unofficial Guide to Paediatrics features an easy-to-read overview of paediatrics, broken down by systems. Each chapter describes core conditions by beginning with aetiology and clinical features, and progresses through investigations, differential diagnoses, management, complications, and finally prognosis. Furthermore, this book covers history taking, examination, communication, and practical skills – all supplemented with clinical cases, labelled diagrams, and information about common examinations and assessment criteria. The authors have also provided illustrations of common procedures and medical devices in clinical practice. As a book produced and written by trainees for other trainees, it captures key information in a digestible manner. With extensive collaboration from renowned academics and specialists, the content is reliable and based on up-to-date evidence.

This textbook is part of an international medical education project, which embodies a passion for peer teaching, and the empowerment of young people who are making a positive impact. Congratulations to Zeshan's team for this award-winning series of textbooks that will help others in their medical journey.

In the wonderful world of paediatrics, this is a wonderful resource for students, junior doctors, and paediatric trainees alike – or anyone looking for a simple and reliable complement to learning from the literature and clinical encounter. The development and success of this has been no child's play – one might even say, it's a milestone of an achievement!

# Abbreviations

4C compound	4-carbon compound	CA-MRSA	Community-associated methicillin resistant	DHEAS	Dehydroepiandrosterone sulphate
5C compound	5-carbon compound		Staphylococcus aureus	DHT	Dihydrotestosterone
17OHP	17-Hydroxyprogesterone		Case-based discussion	DKA	Diabetic ketoacidosis
A	Airway	CBA	Collagen binding activity	DM	Diabetes mellitus
ABG	Arterial blood gas	CBD	Cystathionine beta synthase	DMARD	Disease-modifying antirheumatic drug
ABPA	Allergic bronchopulmonary aspergillosis	CBS	Cognitive behavioural therapy	DMD	Duchenne muscular dystrophy
ACE	Angiotensin-converting enzyme	CBT	Congenital cystic adenomatoid malformation	DMSA	Dimercaptosuccinic acid
ACE	Antegrade colonic enema	CCAM	Crohn's disease	DNA	Deoxyribonucleic acid
Acetyl CoA	Acetyl coenzyme A	CD	Centers for Disease Control and Prevention	DOPA	Dihydroxyphenylalanine
Ach	Acetylcholine	CDC	Congenital dislocation of the hip	DOPS	Direct observation of procedural skills
ACTH	Adrenocorticotrophic hormone	CDH	Constitutional delay of growth and puberty	DRESS	Drug Reaction (or Rash) with Eosinophilia and Systemic Symptoms
ADA	Adenosine deaminase	CDGP	Clinical evaluation exercise	DSD	Disorder of sexual development
ADEM	Acute disseminated encephalomyelitis	CEX	Cystic fibrosis	dsDNA	Double stranded DNA
ADH	Antidiuretic hormone	CF	Cerebral function analysing monitor	DVT	Deep Vein Thrombosis
ADHD	Attention Deficit Hyperactivity Disorder	CFAM	Cystic fibrosis transmembrane conductance regulator	E	Exposure
AED	Anti-epileptic drugs	CFTR	Cystic fibrosis related diabetes	EBV	Epstein-Barr virus
AFP	Alpha fetoprotein	CFRD	Chronic granulomatous disease	ECG	Electrocardiogram
AI	Adrenal insufficiency	CGD	Comparative genomic hybridization	ECMO	Extracorporeal membrane oxygenation
AIDS	Acquired immunodeficiency syndrome	CGH	Coloboma of the eye, heart defect, atresia of the choanae, retarded growth and/or development, genital and/or urinary abnormality, ear abnormality and deafness	ED	Emergency department
AKI	Acute kidney injury	CHARGE syndrome	Congenital heart defect	EDH	Extradural haemorrhage
ALL	Acute lymphoblastic leukaemia	CHD	Conductive hearing loss	EEG	Electroencephalogram
ALP	Alkaline phosphatase	CHL	Creatine kinase	EHEC	Enterohaemorrhagic Escherichia coli
AMH	Anti-Mullerian hormone	CK	Chronic kidney disease	EIEC	Enteroinvasive Escherichia coli
AML	Acute myeloid leukaemia	CKD	Chronic lymphoblastic leukaemia	EMA	Endomyelial antibody
ANA	Antinuclear antibody	CLL	Cow's milk protein	EMG	Electromyography
ANCA	Antineutrophil cytoplasmic antibody	CMP	Cytomegalovirus	ENaC	Epithelial sodium channel
AP	Anteroposterior	CMV	Controlled mandatory ventilation	ENT	Ear, nose and throat
APLS	Advanced Paediatric Life Support	CMV	Central nervous system	EO	Eosinophilic oesophagitis
APS1	Autoimmune polyendocrine syndrome 1	CNS	Carbon dioxide	EPEC	Enteropathogenic Escherichia coli
APTT	Activated partial thromboplastin time	CO <sub>2</sub>	Cerebral palsy	EPLS	European Paediatric Life Support
ART	Antiretroviral therapy	CP	Continuous positive airway pressure	ERT	Enzyme replacement therapy
AS	Aortic stenosis	CPAP	Cardiopulmonary resuscitation	ESR	Erythrocyte sedimentation rate
ASD	Atrial septal defect	CPR	Corticotropin releasing hormone	ESWL	Extracorporeal shock wave lithotripsy
ASD	Autism spectrum disorder	CRH	C-reactive protein	ET	Endotracheal
ASOT	Antistreptolysin O antibody titres	CRP	Cerebrospinal fluid	ETC	Electron transport chain
ATG	Anti-thymocyte globulin	CSF	Computed tomography	ETEC	Enterotoxigenic Escherichia coli
ATP	Adenosine triphosphate	CT	Cardiotocography	EVD	Extraventricular drain
AV	Arteriovenous	CTG	Common variable immunodeficiency	FAH	Fumarylacetoacetase
AVP	Arginine vasopressin	CVID	Central venous line	FBC	Full blood count
AVPU	Alert, voice, pain, unresponsive	CVL	Chorionic villous sampling	FEV <sub>1</sub>	Forced expiratory volume in one second
AVSD	Atrioventricular septal defect	CVS	Chest X-ray	FFP	Fresh frozen plasma
B	Breathing	CXR	Cytochrome P450 enzyme	FGFR	Fibroblast growth factor receptor
BC	Blood culture	D	Disability	Fio <sub>2</sub>	Fraction of inspired oxygen
BCG	Bacillus Calmette-Guérin	DAT	Direct antigen test	FISH	Fluorescence in situ hybridisation
BiPAP	Bi-level Positive Airway Pressure	DDAVP	Desmopressin	fMRI	Functional magnetic resonance imaging
BMA	Bone marrow aspirate	DDH	Developmental dysplasia of the hip	FODMAP	Fermentable Oligo-Di-Mono-saccharides and Polyols
BMD	Becker's muscular dystrophy	DEJ	Dermo-epidermal junction	FPG	Fasting plasma glucose
BMI	Body mass index	DEXA	Dual energy X-ray absorptiometry	FSH	Follicle stimulating hormone
BNF	British National Formulary	DI	Diabetes insipidus	FUO	Fever of unknown origin
BNFc	British National Formulary for Children	DIC	Disseminated intravascular coagulation	FVC	Forced vital capacity
BP	Blood pressure	DIDMOAD	Diabetes insipidus, diabetes mellitus, optic atrophy and deafness	G6PD	Glucose-6-phosphate dehydrogenase
BPM	Beats per minute	DIOS	Distal intestinal obstruction syndrome	GA1	Glutaric aciduria type 1
BTS	British Thoracic Society			GABA	Gamma-aminobutyric acid
BVM	Bag-valve mask			GAD	Glutamic acid decarboxylase
BXO	Balanitis xerotica obliterans			GAGs	Glycosaminoglycans
C	Circulation			GALT	Gut-associated lymphoid tissue
CAFCASS	Children and Family Court Advisory and Support Service			GALT	Galactose-1-phosphate uridylyl transferase
CAH	Congenital adrenal hyperplasia			GARS-2	Gilliam Autism Rating Scale
CAKUT	Congenital anomalies of the kidney and urinary tract				
CAMHS	Child and Adolescent Mental Health Service				

GAVI	Global Alliance for Vaccines and Immunisation	ITP	Idiopathic thrombocytopenic purpura	OGTT	Oral glucose tolerance test
GBM	Glomerular basement membrane	IUD	Intrauterine device	OME	Otitis media with effusion
GBS	Group B streptococcus	IUGR	Intrauterine growth restriction	OP	Oropharyngeal
GBS	Guillain-Barré syndrome	IUS	Intrauterine system	ORS	Oral rehydration solution
GCS	Glasgow coma scale	IV	Intravenous	OSCE	Objective structured clinical examination
GFR	Glomerular filtration rate	IVA	Isovaleric acidaemia	OT	Occupational therapy
GH	Growth hormone	IVH	Intraventricular haemorrhage	PA	Propionic acidaemia
GHD	Growth hormone deficiency	IVIG	Intravenous immunoglobulin	PALS	Patient advice and liaison service
GLUT-1	Glucose transporter 1	JIA	Juvenile idiopathic arthritis	PANDAS	Paediatric autoimmune neuropsychiatric disorders associated with streptococcal infections
GOR	Gastro-oesophageal reflux	JVP	Jugular venous pressure	PAPP-A	Pregnancy-associated plasma protein-A
GORD	Gastro-oesophageal reflux disease	KDOQI	Kidney Disease Outcomes Quality Initiative	PCD	Primary ciliary dyskinesia
GP	General practitioner	KUB	Kidney-ureter-bladder	PCDAI	Paediatric Crohn's Disease Activity Index
GnRH	Gonadotropin-releasing hormone	LARC	Long acting reversible contraception	PCP	Pneumocystis carinii (jirovecii) pneumonia
GSDs	Glycogen storage disorders	LCH	Langerhans cell histiocytosis	PCR	Polymerase chain reaction
GvHD	Graft-versus-host disease	LDH	Lactate dehydrogenase	PCV	Packed cell volume
HAART	Highly active antiretroviral therapy	LFT	Liver function test	PDA	Patent ductus arteriosus
Hb	Haemoglobin	LH	Luteinising hormone	PE	Pulmonary Embolism
HbA1c	Glycated haemoglobin	LP	Lumbar puncture	PEEP	Positive end expiratory pressure
HbS	Sickle cell haemoglobin	LTOT	Long term oxygen therapy	PEF	Peak expiratory flow
HcG	Human chorionic gonadotropin	MAG3	Mercurioacetyltriglycine	PEG	Percutaneous endoscopic gastrostomy
HD	Hodgkin's disease	MAP	Mean arterial pressure	PEG-J	Percutaneous endoscopic gastro-jejunostomy
HDU	High dependency unit	MAP	Mean airway pressure	PET	Positron emission tomography
HFNC	High flow nasal cannulae	MCAD	Medium-chain acyl-CoA dehydrogenase	PG	Plasma glucose
HFOV	High-frequency oscillatory ventilation	MCADD	Medium-chain acyl-CoA dehydrogenase deficiency	PHEX	Phosphate regulating endopeptidase homolog
HH	Hypogonadotropic hypogonadism	MC&S	Microscopy, culture and sensitivity	PICC	Peripherally inserted central catheter
HiB	Haemophilus influenza type B	MCUG	Micturating cystourethrogram	PICU	Paediatric intensive care unit
HIE	Hypoxic-ischaemic encephalopathy	MCV	Mean cell volume	PIP	Peak inspiratory pressure
HIV	Human Immunodeficiency Virus	MDGs	Millennium Development Goals	PKU	Phenylketonuria
HLA	Human leucocyte antigen	MDI	Metered-dose inhaler	PMTCT	Prevention of mother-to-child transmission
HLH	Haemophagocytic lymphohistiocytosis	MDR	Multi-drug resistant	PNET	Primitive neuroectodermal tumour
HPA	Hypothalamic-pituitary-adrenal	MDT	Multidisciplinary team	PORCN	Porcupine homolog
HPV	Human papilloma virus	MEN	Multiple endocrine neoplasia	PP	Precocious puberty
HPG	Hypothalamic-pituitary-gonadal	MenC	Meningococcus C	PPHN	Persistent pulmonary hypertension of the newborn
HR	Heart rate	MG	Myasthenia gravis	PPV	Patent processus vaginalis
HRCT	High resolution computed tomography	MIBG	Metaiodobenzylguanidine	PR	Per rectum
HS	Hereditary spherocytosis	MIS	Müllerian-inhibiting substance	PRO	Parental Responsibility Order
HSCT	Haemopoietic stem cell transplantation	MMA	Methylmalonic acidaemia	PROM	Premature rupture of the membranes
HSP	Henoch Schönlein Purpura	MMR	Measles, mumps and rubella	PRN	Pro re nata i.e. as required
HSV	Herpes simplex virus	MODY	Maturity onset diabetes of the young	PS	Pulmonary stenosis
HUS	Haemolytic uraemic syndrome	MPS	Mucopolysaccharidoses	PT	Prothrombin time
HVA	Homovanillic acid	MRCPC	Membership of the Royal College of Paediatrics and Child Health	PTV	Patient triggered ventilation
IBD	Inflammatory bowel disease	MRI	Magnetic resonance imaging	PTH	Parathyroid hormone
IBS	Irritable bowel syndrome	MRSA	Methicillin resistant	PT	Physiotherapy
IC	Indeterminate colitis	MSUD	Staphylococcus aureus	PTSD	Post-traumatic stress disorder
ICD	Implantable cardioverter defibrillator	NAD	Maple syrup urine disease	PUCAI	Paediatric Ulcerative Colitis Activity Index
ICP	Intracranial pressure	NAI	Nicotinamide adenine dinucleotide	PUJ	Pelvi-ureteric junction
ICU	Intensive care unit	NBM	Non-accidental injury	PUV	Posterior urethral valve
Ig	Immunoglobulin	NEC	Nil by mouth	PVL	Periventricular leukomalacia
IgA	Immunoglobulin A	NF	Necrotising enterocolitis	PWS	Prader-Willi syndrome
IGF1	Insulin-like growth factor 1	NF1/2	Neurofibromatosis	RBC	Red blood cell
IGF2	Insulin-like growth factor 2	NG	Neurofibromatosis type 1 or 2	REM	Rapid eye movement
IL-2	Interleukin-2	NHL	Nasogastric	ReSoMal	Rehydration solution for malnutrition
IL-4	Interleukin-4	NICE	Non-Hodgkin lymphoma	RF	Rheumatoid factor
IL-5	Interleukin-5	NKF	National Institute for Health and Care Excellence	RICOF	Ristocetin cofactor
IL-13	Interleukin-13	NLS	Neonatal intensive care unit	RIF	Right iliac fossa
ILAE	International League Against Epilepsy	NMDA	Newborn and infant physical examination	RNA	Ribonucleic acid
IIH	Idiopathic Intracranial hypertension	NPA	Non-Invasive Positive Pressure Ventilation	RR	Respiratory rate
IM	Intramuscular	NSAIDS	National Kidney Foundation Newborn life support	RSV	Respiratory syncytial virus
INR	International normalised ratio	NSPCC	N-methyl-D-aspartate	RUTF	Ready-to-use therapeutic food
IO	Intraosseous	NTD	Nasopharyngeal aspirate	SAH	Subarachnoid haemorrhage
IPS	Infantile pyloric stenosis	O <sub>2</sub>	Non-steroidal anti-inflammatory drugs	SALT	Speech and language therapy
IQ	Intelligence quotient	OA	National Society for the Prevention of Cruelty to Children	SBAR	Situation, background, assessment and recommendation
IRT	Immunoreactive trypsinogen	OAE	Neural tube defect	SC	Serum bilirubin
ISS	Idiopathic short stature		Oxygen		Sickle cell anaemia
ITP	Immune thrombocytopenia		Osteoarthritis		
			Oto-acoustic emissions		

SCAD	Short-chain acyl-CoA dehydrogenase	SUFE	Slipped upper femoral epiphysis	UAC	Umbilical artery catheter
SCBU	Special care baby unit	SVCO	Superior vena cava obstruction	UC	Ulcerative colitis
SCID	Severe combined immunodeficiency	SVD	Spontaneous vaginal delivery	UDT	Undescended testes
SD	Standard deviation	SVT	Supraventricular tachycardia	U&E	Urea and electrolytes
SDGs	Sustainable Development Goals	SWAN	Syndrome without a name	UO	Ureteric orifice
SDH	Subdural haemorrhage	SWS	Sturge-Weber syndrome	UN	United Nations
SEDU	Specialist eating disorders unit	T1DM	Type 1 diabetes mellitus	URTI	Upper respiratory tract infection
SENCO	Special Educational Needs Coordinator	T2DM	Type 2 diabetes mellitus	USS	Ultrasound scan
SGA	Small for gestational age	Te	Expiratory Time	UTI	Urinary tract infection
SHOX	Short Stature Homeobox gene	Ti	Inspiratory Time	UV	Ultraviolet
SIADH	Syndrome of inappropriate antidiuretic hormone secretion	TAC	Team around the child	UVC	Umbilical vein catheter
SIDS	Sudden infant death syndrome	TAPVD	Total anomalous pulmonary venous drainage	VACTERL association	Vertebral defects, anal atresia, cardiac defects, tracheo-oesophageal fistula, renal anomalies and limb abnormalities
SIGN	Scottish Intercollegiate Guidelines Network	TB	Tuberculosis	VHL	Von Hippel-Lindau disease
SIMV	Synchronised intermittent mandatory ventilation	TCA	Tricarboxylic acid	VLCAD	Very-long-chain acyl-CoA dehydrogenase
SLE	Systemic lupus erythematosus	TDS	Ter die sumendum ie. Three times a day	VMA	Vanillylmandelic acid
SMA	Spinal muscular atrophy	TFT	Thyroid function test	VMAT	Vesicular monoamine transporter
SNHL	Sensorineural hearing loss	Tg	Thyroglobulin	VP	Ventriculoperitoneal
SPECT	Single-photon emission computed tomography	tTGA	Tissue transglutaminase antibody	VSD	Ventricular septal defect
SpO2	Oxygen saturation	Th2	Type 2 Helper T-Cell	VT	Ventricular tachycardia
SROM	Spontaneous rupture of membranes	Thal	Thalassaemia	VTEC	Verotoxin producing Escherichia coli
SRS	Social Responsiveness Scale	TIA	Transient ischaemic attack	VUJ	Vesicoureteric junction
SSPE	Subacute sclerosing panencephalitis	TIBC	Total iron binding capacity	VUR	Vesico-ureteric reflux
SSRI	Selective serotonin reuptake inhibitor	TLS	Tumour lysis syndrome	VZV	Varicella zoster virus
STEC	Shiga Toxin-Producing E. coli	TNF	Tumour necrosis factor	WAS	Wiskott-Aldrich syndrome
SUDEP	Sudden unexpected death in epilepsy	TORCH	Toxoplasmosis, Other, Rubella, CMV, Herpes	vWD	von Willebrand disease
SUDI	Sudden unexpected death in infancy	TPMT	Thiopurine methyltransferase	vWF	von Willebrand factor
		TPN	Total parenteral nutrition	WCC	White cell count
		TRH	Thyrotropin-releasing hormone	WHO	World Health Organisation
		TSH	Thyroid-stimulating hormone		
		TST	Tuberculin skin test		
		tTG	Tissue transglutaminase		
		TTN	Transient tachypnoea of the newborn		

# Contributors

## Chief Editor



**Zeshan Qureshi** BM MSc BSc (Hons) MRCPCH  
Academic Clinical Fellow, Great Ormond  
Street, and Institute of Global Health, UCL, UK

## Associate Editor



**Tina Sajjianhar** MBBS DRCOG DCH FRCPCH  
FCEM  
Consultant in Paediatric Emergency  
Medicine, Divisional Director for Children  
and Young People services, Lewisham and  
Greenwich NHS Trust, UK

## Authors



**John Jungpa Park** MB ChB MTh  
BMedSci (Hons)  
Junior Doctor, North Central Thames  
Foundation School, UK



**Maxine Wilke**  
Medical Student, Keele University, UK



**Chi Hau Tan** MBBS (Hons) GDipSurgAnat  
Neurosurgery Registrar, Monash Health,  
Melbourne, Australia



**Anna Capsomidis** BSc MBChB (Hons)  
MRCPCH PGDip (Med Ed) PGC (Healthcare  
Ethics and Law)  
Clinical Research Fellow, UCL, Great Ormond  
Street Institute of Child Health, UK



**Anand Goomany** MbChB BSc  
Core Surgical Trainee, Bradford Teaching  
Hospitals Foundation Trust, West  
Yorkshire, UK



**May Bisharat** MBBS (Hons) MSc FRCS  
(Paed Surg)  
Registrar in Paediatric Surgery, Evelina  
Children's Hospital, London, UK



**Amy Mitchell** MBChB BSc (Hons) MSc  
DipClinEd MRCPCH  
Consultant Paediatric Oncologist,  
Southampton University Hospital Trust,  
Southampton, UK



**Rachael Mitchell** MRCPCH MA (Cantab)  
Paediatric Registrar, London Deanery,  
London, UK



**David K K Ho** MBChB DTM&H MRCPCH  
Clinical Research Fellow, Institute of Child  
Health, University College London, UK



**Alexander Young** MBChB MSc MRCS PGCME  
Trauma and Orthopaedic Surgery Registrar,  
Severn Deanery, UK



**Christopher Harris** MBChB MRCPCH  
Paediatric Neonatal Registrar, St Georges  
Hospital, London, UK



**Marie Monaghan** MBBS BSc (Hons) MRCPCH  
Paediatric Registrar, London Deanery, UK



**Marylyn-Jane Emedo** MBBS BSc MRCPCH  
Paediatric Registrar, London Deanery,  
London, UK



**Antonia Hargadon-Lowe** BMBS BMedSci  
MRCPCH PGC (Paed)  
Paediatric Registrar, London Deanery,  
London, UK



**Sadhanandham Punniyakodi** MBBS MRCPCH MSc  
Senior Training Fellow in Paediatric Endocrinology, The Great North Children's Hospital, Royal Victoria Infirmary, Newcastle upon Tyne



**Stephen D Marks** MD MSc MRCP DCH FRCPCH  
Consultant Paediatric Nephrologist, Great Ormond Street Hospital for Children NHS Foundation Trust, London, UK.



**Hannah Linford** BMBS MRCPCH  
Paediatric Registrar, KSS Deanery, UK



**Anita Demetriou** MBBS BSc MRCPCH DTMH  
Paediatric Registrar, London Deanery, UK



**Christopher John Grime** MBChB MRCPCH  
Paediatric Registrar, London Deanery, UK.



**Debasree Das** MBBS BSc MRCPCH  
Paediatric Registrar, London Deanery, UK



**Philippa King** BSc MBChB MRCPCH MSc  
Academic Clinical Fellow, Medical Microbiology, East of England Deanery, UK



**Isabel Mawson** MBBS BSc MRCPCH  
Paediatric Registrar, London Deanery, UK



**Claire Bryant** BMedSc BMBS  
Junior Doctor, South Thames Foundation School, London, UK



**Anna Chadwick** MBBS BSc MRCPCH  
Paediatric Registrar, London Deanery, UK



**Andy Hall** MBChB MRCS DOHNS  
ENT Specialist Registrar, Great Ormond Street, UK



**Amy Moran** MBBS MRCPCH  
Paediatric Registrar, London Deanery, UK



**Michael Malley** MA (Hons) Cantab MBBS MRCPCH  
Paediatric Registrar, London Deanery, UK



**Alice Armitage** MBBS BSc MRCPCH  
Academic Clinical Fellow, Paediatrics, London Deanery, UK



**Vaitza Tziaferi** MD MRCPCH MSc  
Consultant in Paediatric Endocrinology & Diabetes, Leicester Royal Infirmary, UK



**Kunal Babla** BSc(Hons) MBBS MSc MRCPCH MAcadMEd  
Neonatal Registrar, London Deanery, UK



**Maanasa Polubothu** BSc MBChB MSc MRCPCH PGDip (Genomic Medicine)  
Academic Clinical Fellow, London Deanery, UK



**Sam Thenabadu** MBBS MRCP DRCOG DCH MA Clin Ed FCEM MSc (Paed) FHEA  
Consultant Adult & Paediatric Emergency Medicine. Honorary Senior Lecturer, King's College London.



**Stephanie Connaire**

Final Year Medical Student, Cardiff University, Cardiff, UK



**Zainab Kazmi** BSc (Hons) MBChB

Academic Foundation Doctor and Honorary Clinical Fellow, University of Glasgow



**Pooja Parekh** MBBS

Paediatric Trainee, London Deanery, UK

**Expert Reviewers**

**Dr Vandy Bharadwaj**

Consultant Paediatric Haematologist, Southampton Children’s Hospital, Southampton, UK

**Prof. Robert Tulloh** MA DM FRCP FRCPCH

Consultant Paediatric Cardiologist, Bristol Royal Hospital for Children, UK. Honorary Professor of Congenital Cardiology and Pulmonary Hypertension, University of Bristol

**Prof. David Walker**

Professor of Paediatric Oncology, Children’s Brain Tumour Research Centre, Nottingham, UK

**Mr RA Wheeler** MS FRCS LLB (Hons) LLM

Consultant Neonatal & Paediatric Surgeon. Director, Department of Clinical Law. University Hospital of Southampton, UK.

**Mr Theo Joseph** FRCS

ENT Consultant, Royal National Throat Nose and Ear Hospital, London, UK

**Dr Victoria Jones** MRCPCH

Consultant Paediatrician, North Middlesex University Hospital, UK

**Dr Anne-Marie Ebdon** MBBS MRCPCH FRACP

Consultant Paediatrician, Queen Mary’s Hospital for Children, Epsom & St.Hellier University Hospitals NHS Trust, UK

**Dr Sarah K Clegg** MBChB BSc MRCP UK MRCPCH

Consultant Paediatrician, Department of Community Child Health, Edinburgh, UK

**Dr Solomon Kamal Uddin** MBBS BMedSci

Paediatric Registrar, London Deanery, UK

**Dr Delan Devakumar** MBChB MSc MRCPCH DTM&H

MFPH PhD  
Public Health Registrar, London Deanery, UK

**Dr Khadija H Aljefri** MBChB, MRCP (UK), MRCP (Derm)

Dermatology Registrar, Royal Victoria Infirmary, Newcastle upon Tyne, UK

**Mr Neil Tickner** MRPharmS PGDip. Clin. Pharm

Lead Pharmacist Paediatrics, St Mary’s Hospital, London, UK

**Stephanie Connaire**

Final Year Medical Student, Cardiff University, Cardiff, UK

**Dr Pooja Parekh** MBBS

Paediatric Trainee, London Deanery, UK

**Bianca Davis**

School Teacher, Head of Personal Social and Health Education, London, UK

**Dr Sreena Das** MB ChB MRCPCH

Consultant Paediatrician, King’s College Hospital, London, UK

**Dr Simon Chapman** BA BM FRCPCH

Consultant Paediatrician, King’s College Hospital, London, UK

**Dr Isabel Farmer** MBBS MRCPCH

Haematology Registrar, Queen Elizabeth Hospital, London, UK

**Tabatha English** BSc (Hons) RM

Midwife, Lewisham Hospital, London, UK

**Kelly Frogbrook** PgDip RN

Paediatric Nurse, Queen Elizabeth Hospital, London, UK

**Dan Purnell**

Lead Resuscitation Officer, Lewisham and Greenwich Healthcare NHS Trust

**Dr Daniel Langer** BSc (Hons) MRCPCH DPID

Consultant in Acute & Ambulatory Paediatrics. Special interest in paediatric infectious diseases (SPIN ID) and global child health  
Epsom & St Helier NHS Trust, UK

**Lydia Shackshaft**

Medical Student, King’s College London, UK

**Sammie Mak**

Medical Student, Leeds University, UK

**Ben Evans**

Medical Student, University of Newcastle, UK

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# 1.01

## ADOLESCENT MEDICINE

ALICE ARMITAGE

### CONTENTS

- 1 Approach to the Adolescent Consultation**
  - 1 Adolescent History Taking
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  - 2 Puberty, Growth and Development
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- 6 Transition from Paediatric to Adult Services**
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### APPROACH TO THE ADOLESCENT CONSULTATION

**A**dolescence is the transitional phase of growth and development between childhood and adulthood. An adolescent is defined by the World Health Organisation as a person between 10 and 19 years of age. There is increasing recognition of the specific problems of this age group, including trauma, mental health issues, pregnancy and sexually transmitted diseases. The rise in healthcare usage in adolescence is multifactorial; increasing survival from chronic childhood conditions, use of drugs and alcohol, and risk-taking behaviour all play a role, and advances in perinatal care and immunisation have shifted the burden of disease away from the under-fives. Understanding the unique needs of this age group is a core skill for any physician. The law relating to children and adolescents is covered in Chapter 1.13.

#### Adolescent History Taking

It is important to adapt the approach to the needs of the adolescent age-group. At the start of a consultation, consider the following:

- ▶ Always speak to the patient, not to their parent or carer, unless there is no alternative.
- ▶ Ask the patient if they would like to speak to alone or ask them if they would like someone to be present, such as a parent or friend.
- ▶ For any examination, offer a chaperone.
- ▶ Talk in an age-appropriate fashion. It is easy to alienate an adolescent patient by appearing patronising or by using medical jargon.
- ▶ Try to anticipate issues around consent and confidentiality.

The psychosocial history is vital to the adolescent history. Many presentations will stem from an issue like drug use, a fight with a partner or worries about sexuality. A useful aide-memoire is the HEADSS tool, shown in *Table 1*. In general, start with more open questions and then focus questions to the information given. A common reason for missing important issues is making assumptions; for example, thinking that all young people live at home with their parents or that all young people are heterosexual.





# 1.02

## ASSESSMENT AND MANAGEMENT OF THE ACUTELY UNWELL CHILD

CHRISTOPHER HARRIS

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- 15 The Death of an Infant or Child**
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  - 16 Debriefing
- 17 References and Further Reading**

### INTRODUCTION

Clinicians are fearful of treating children and infants in the Emergency Department (ED), and, more generally, of making medical decisions concerning children. Junior doctors manage children in many settings (*Box 1*).

#### Box 1: Settings in which unwell children may be encountered

- Primary Care.
- Emergency Department.
- Paediatric Emergency Department.
- Paediatric wards.
- Adult specialties with paediatric cover (surgery, orthopaedics, ENT and dermatology).

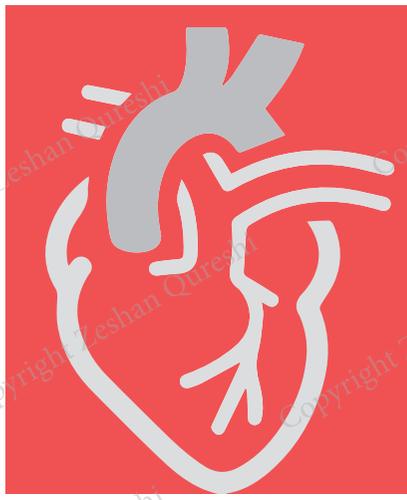
This chapter provides a framework for assessing any child that presents for medical attention to ensure they receive timely and effective care, with a particular focus on dealing with the very sick child.

When approaching a potentially sick child, bear in mind that children are not just small adults. Different challenges and techniques must be addressed to get to the bottom of the problem quickly.

Consider the following:

- ▶ The parents or carer may not have the whole story if the child has been in the care of someone else, and so a collateral history may be necessary.
- ▶ The parents' "sixth sense" regarding their child should always be taken seriously, but equally, parents may underestimate the seriousness of their child's condition.
- ▶ Younger children may not be able to give any history, or the history given may be misleading.
- ▶ All children, but particularly younger infants, may have underlying congenital abnormalities that have remained undetected until presentation in extremis.





# 1.03

## CARDIOLOGY

HANNAH LINFORD



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### CARDIAC RHYTHMS

#### Normal Cardiorespiratory Parameters

In children, the normal range for heart rate, blood pressure and respiratory rate vary significantly with age. *Table 1* summarises these features.

#### The Paediatric ECG

Interpreting an electrocardiogram (ECG) in children is not the same as with an adult because the heart is physiologically different. Points to note are:

- ▶ **Normal heart rate is much higher.** A heart rate of over 150 bpm is normal in a newborn. Conversely, for a newborn, a heart rate less than 100 needs further assessment.
- ▶ **Normal marked sinus arrhythmia.** This is because the heart rate varies dramatically with breathing.
- ▶ **Cardiac axis is deviated to the right in the newborn.** This is because there is marked right ventricular hypertrophy, as the right ventricle pumps blood against a high-resistance collapsed lung in utero. Often, the right ventricle is captured by placing an additional lead (V4R) in the fifth intercostal space, at the mid clavicular line on the right. V1 (looking directly at the right ventricle) and V2-V3 often have a dominant R wave.
- ▶ **Possible partial right bundle branch block.** This manifests as a normal QRS complex, plus an RSR pattern (M shape) in V1.
- ▶ **T wave inversion.** This is normal in leads V1-3 and potentially V4 as well.
- ▶ **Q waves.** These are normal in the inferior (AVF, II, III) and left precordial leads (V5-6).

The normal ranges for ECG interpretation change with age. *Table 2* and *Table 3* show the QRS complex and corrected QT interval. *Figure 1* shows a normal ECG and *Figure 2* shows the calculation of the QT interval.

**TABLE 1: Normal values for heart rate, blood pressure and respiratory rate in children. Source: Advanced Paediatric Life Support, the practical approach. Fifth edition – Advanced Life support group.**

Normal ranges of routine observations in children.				
	<1 year.	2-5 years.	5-12 years.	12-18 years.
Heart rate.	110-160bpm.	95-140bpm.	80-120bpm.	60-100bpm.
Systolic blood pressure (50 <sup>th</sup> centile).	80-90mmHg.	85-100mmHg.	90-110mmHg.	90-120mmHg.
Respiratory rate.	30-40 breaths/min.	25-30 breaths/min.	20-25 breaths/min.	15-20 breaths/min.

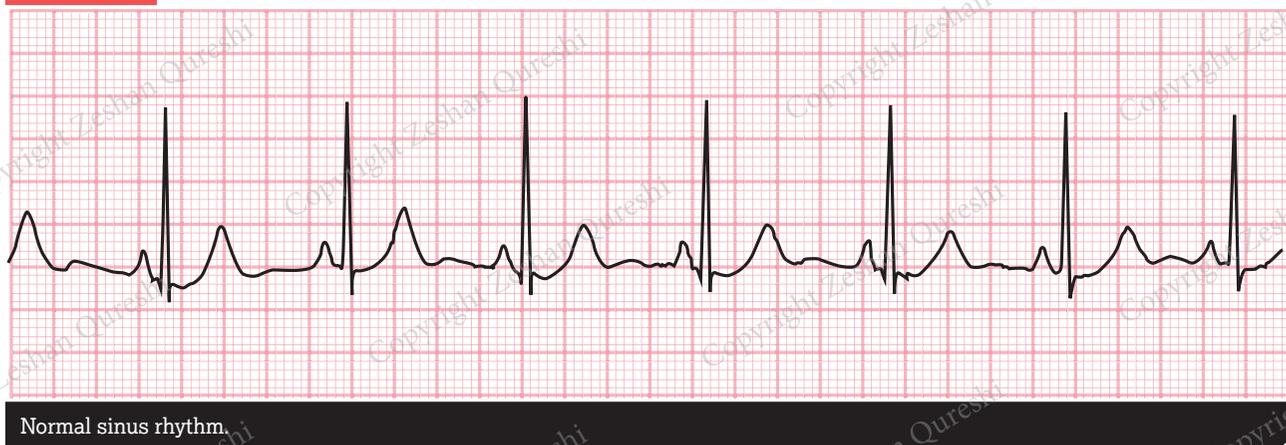
**TABLE 2: QRS ranges in children**

Age	Cardiac Axis
1 week – 1 month.	+ 110° (range +30° to +180°).
1 month – 3 months.	+ 70° (range +10° to +125°).
3 months – 3 years.	+ 60° (range +10° to +110°).
Over 3 years.	+ 60° (range +20° to +120°).
Adult.	+ 50° (range -30° to 105°).

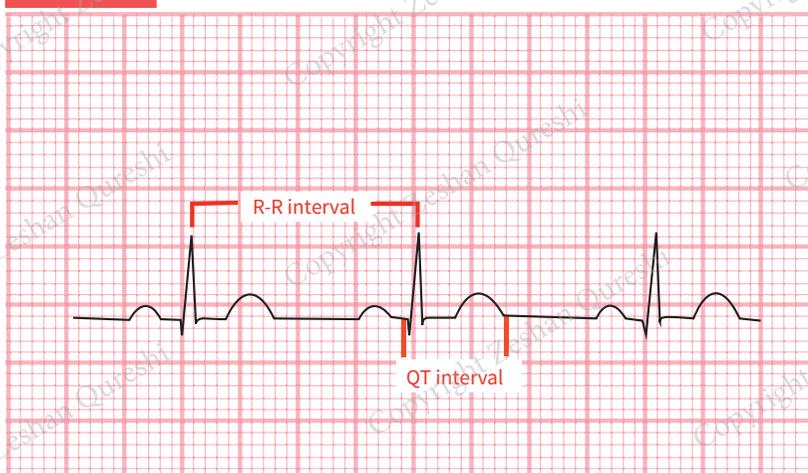
**TABLE 3: Corrected QT interval**

Age	QT Interval
< 6 months.	<0.49 seconds.
>6 months.	<0.44 seconds.

**FIGURE 1**



**FIGURE 2**



Location of the QT interval, from beginning of Q to end of T in the same cycle. Note that this needs to be compared to the preceding RR interval to get the corrected QT interval (QT/√RR).

**Tachycardia**

Tachycardia may result from physiological processes that alter the sympathetic/parasympathetic tone, resulting in a sinus tachycardia. Such causes include:

- Excessive activity.
- Crying/being upset.
- Stress.

It may also be caused by a secondary problem outside the heart, like fever, infection, hyperthyroidism, anaemia or any other problem causing a high metabolic rate. It is rarely due to a primary cardiac cause. A primary cause is more likely if tachycardia is particularly high or an isolated finding.



TABLE 6: Continued

Disease	Definition	Presentation	Management
Hypoplastic left heart syndrome.	A group of defects affecting the left side of the heart (valves and chambers) in which the structures are too small to support systemic output.	<ul style="list-style-type: none"> <li>This is usually antenatally diagnosed – with an aim for delivery at a cardiac centre.</li> <li>It presents as a breathless, severely cyanotic and compromised neonate soon after birth.</li> <li>ECG shows absent ventricular forces.</li> </ul>	<ul style="list-style-type: none"> <li>A prostaglandin infusion is initially started to maintain duct patency.</li> <li>Urgent transfer to a specialist cardiac unit is needed.</li> <li>There is a three stage surgical treatment (e.g. Norwood, Glenn, then Fontan). The ultimate aim is for the right ventricle to remain the systemic ventricle, with blood passively flowing to the lungs. Heart transplant is another option.</li> </ul>
Tricuspid atresia.	The tricuspid valve (between the right atrium and right ventricle) is blocked or absent. Therefore, deoxygenated blood cannot flow into the pulmonary circulation from the right ventricle. This defect is incompatible with life unless it is accompanied by both a VSD and an ASD or a PDA.	<ul style="list-style-type: none"> <li>Cyanosis is present soon after birth.</li> <li>A systolic murmur is heard at the left lower sternal edge if a VSD is present or a continuous murmur below the left clavicle if a PDA is present.</li> </ul>	<ul style="list-style-type: none"> <li>Surgical correction occurs by a course of several staged procedures. Initial palliation may involve inserting a systemic-pulmonary shunt (e.g. Blalock-Taussig shunt).</li> <li>Systemic venous return is connected to the pulmonary artery, bypassing the right ventricle (Fontan circulation).</li> </ul>
Complete atrioventricular septal defect (AVSD).	A large ASD which is continuous with a large VSD; all four chambers of the heart are communicating with one another. This causes increased blood flow to the lungs as well as less effective drainage, thereby promoting pulmonary hypertension and heart failure.  Note that smaller AVSDs may be acyanotic.	<ul style="list-style-type: none"> <li>Heart failure and cyanosis.</li> <li>This is the abnormality most often associated with Down syndrome.</li> <li>Ejection systolic murmur at left upper sternal edge (pulmonary flow murmur).</li> <li>Pansystolic murmur at the apex (mitral regurgitation).</li> <li>Fixed splitting S2 (ASD).</li> <li>Loud S2 (pulmonary hypertension).</li> </ul>	<ul style="list-style-type: none"> <li>The heart failure is medically managed.</li> <li>The VSD and ASD are surgically closed and the atrioventricular valve is surgically repaired (to fashion separate mitral and tricuspid valves).</li> </ul>

FIGURE 8

Patent ductus arteriosus

Atrial septal defect

Transposition of great arteries

■ Oxygen rich blood  
■ Oxygen poor blood  
■ Mixed blood

AO = Aorta  
 PA = Pulmonary artery  
 LA = Left atrium  
 RA = Right atrium  
 LV = Left ventricle  
 RV = Right atrium

Transposition of the great arteries.

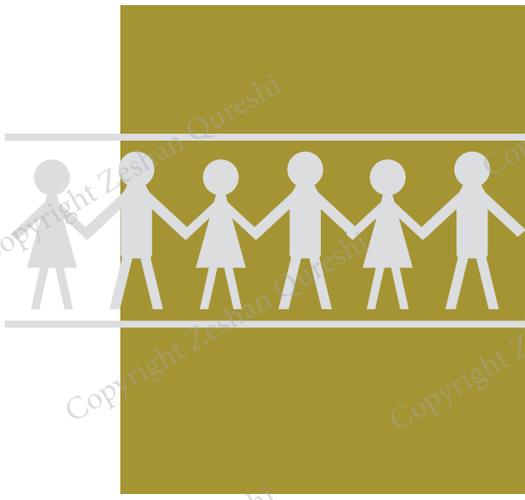
FIGURE 9

Ventricular septal defect

■ Oxygen rich blood  
■ Oxygen poor blood  
■ Mixed blood

LA = Left atrium  
 RA = Right atrium  
 LV = Left ventricle  
 RV = Right atrium

Ventricular Septal Defect.



# 1.04

## COMMUNITY PAEDIATRICS

ANTONIA HARGADON-LOWE

### DEVELOPMENT

#### Normal Development and the Developmental Assessment

Developmental history is often missed when seeing children in acute settings or in primary care. This may be due to time constraints in a busy Emergency Department (ED) or outpatient clinic. However, some information can be gathered quickly with adequate knowledge and practice. Picking up developmental problems early can have a significant impact on that child's outcome. The time taken and depth of questioning on development depends on the clinical scenario (*Table 1*).

TABLE 1: Developmental assessment in different settings

Scenario	Suggested Approach
3-year-old child presenting to ED with a fever and ear pain, and no known developmental concern.	Brief questioning about milestones and parental concerns, e.g. <ul style="list-style-type: none"> <li>Do you feel your child is able to do the same things other 3-year-olds can?</li> <li>At what age did she smile, say her first word and walk?</li> </ul>
Outpatient clinic review of 2-year-old boy who has had three seizures in the last few months.	More in-depth questioning is required as relevant to the presenting complaint. The number of questions will depend on the answers and whether concerns emerge. Ascertain whether he is similar to other children of his age. Developmental regression should also be enquired about, i.e. has he lost any skills previously developed. To start, ask a couple of questions in each developmental domain to assess whether he is meeting expected targets for his age: <ul style="list-style-type: none"> <li><b>Gross motor.</b> Is he able to jump? Is he able to climb stairs, and if so, how does he do it?</li> <li><b>Fine motor.</b> Can he draw a straight line? How big a tower block can he build with building blocks?</li> <li><b>Language/hearing.</b> Can he speak in short sentences? Can he obey simple commands?</li> <li><b>Social/Self-Care.</b> Can he eat with a fork/spoon? Does he socially interact (good eye contact and smiles) with adults and other children?</li> <li><b>General.</b> Has he lost any skills that he could previously do?</li> </ul>
Community paediatric outpatient appointment following a referral from nursery school with concerns of development.	In this situation, the reason for the referral is "developmental concerns", so a full assessment taking approximately one hour is necessary using one of the available tools.

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#### Developmental Milestones

Development is usually categorised into distinct areas; commonly the following domains are used: "Gross Motor", "Fine Motor/Vision", "Language" and "Social/Self-Care" (*Table 2*). This is done because developmental milestones can often be grouped together, and if one sector is affected but not the others, it may indicate a particular group of pathologies. For example, language delay may be secondary to hearing loss, whereas hearing loss is unlikely to cause isolated gross motor delay.



SAFEGUARDING

Categories of Abuse

Child protection is relevant in every aspect of paediatrics, and more generally for anyone dealing with children. According to the National Society for the Prevention of Cruelty to Children (NSPCC), in the UK approximately two children die per week from homicide, neglect or “undetermined intent”.

Abuse can be divided into four categories:

- ▶ Physical abuse.
- ▶ Neglect.
- ▶ Sexual abuse.
- ▶ Emotional abuse.

These are not mutually exclusive and often co-exist.

Physical Abuse

Physical abuse encompasses any physical harm caused to a child. **Figure 3** shows the typical sites of accidental and non-accidental injury (NAI). **Figure 4** shows specific non-accidental injuries. This may involve hitting, biting, burning or scalding, shaking, throwing, poisoning, suffocation, drowning, or any cause of physical harm to a child. It includes fabricated or induced illness. Note:

- ▶ These injuries can present as bruises, burns, lacerations, bite marks, petechiae, fractures or neurological signs suggesting intracranial injuries.
- ▶ Specific patterns of bruising are more suspicious than others:
  - › Marks that are well demarcated, have the appearance of a specific implement (such as a belt), or look like finger marks from hitting or gripping.
  - › Bruises that are not on bony prominences (inner/posterior thighs, buttocks, trunk, chest, behind or on ears, neck or shoulders, intra-oral), or bilateral.
- ▶ Bruises cannot be aged accurately.
- ▶ In burns, look for splash marks and irregular edges to support a story of accidental burns.
- ▶ There should be a strong suspicion of NAI in spiral, oblique or metaphyseal fractures, but a consistent and good story could explain these, particularly in older children.
- ▶ NAI should be strongly suspected and actively excluded in any non-mobile child or child under 1-year-old with a fracture unless there is a very plausible mechanism of injury.

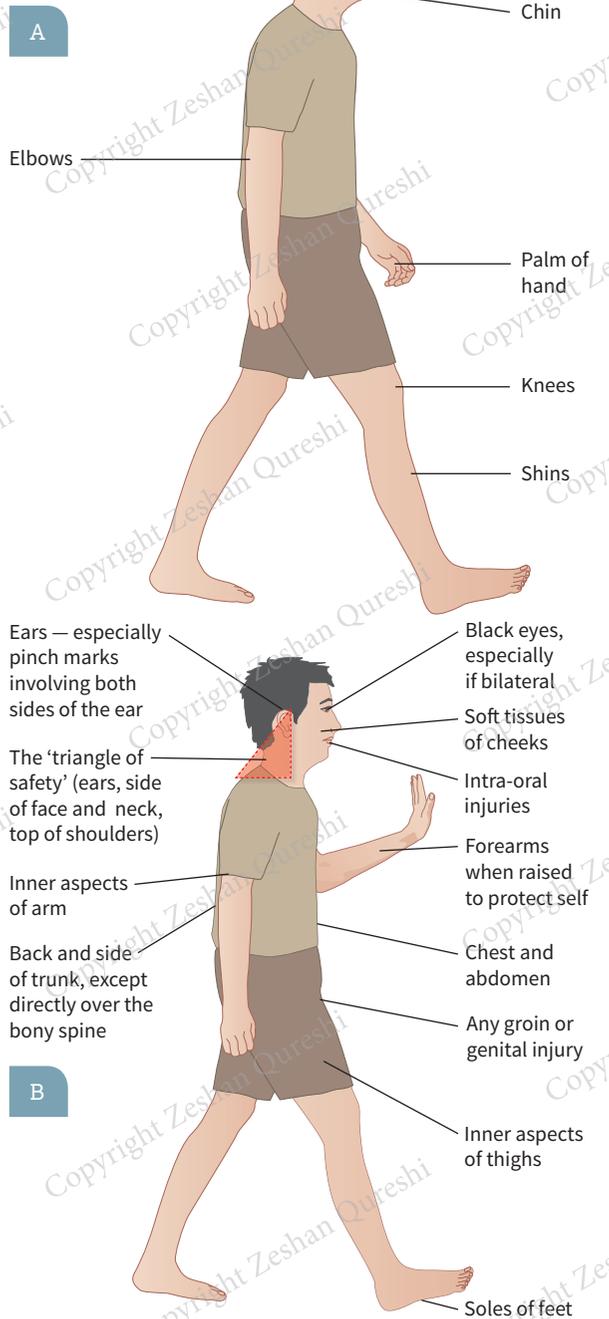
Neglect

This is the persistent failure to meet a child’s basic physiological and psychological needs and has a negative impact on their health and development. It can include:

- ▶ Poor diet.
- ▶ Meeting health needs.
- ▶ Educational achievement.
- ▶ Hygiene.
- ▶ Protection from danger and prevention of harm.

FIGURE 3

Head injuries tend to involve the parietal bone, occiput and forehead



Typical sites of A) accidental injury and B) non-accidental injury.

- ▶ Protection from witnessing violence.
- ▶ Provision of emotional warmth.

Sometimes, neglect is the only concern preceding a child death from abuse, and therefore the significance of neglect should not be underestimated. Every paediatrician assessing children should be alert to signs of neglect of a child’s welfare. One of the best ways to chart wellbeing is through growth parameters,





# 1.05

## ENT

ANDY HALL



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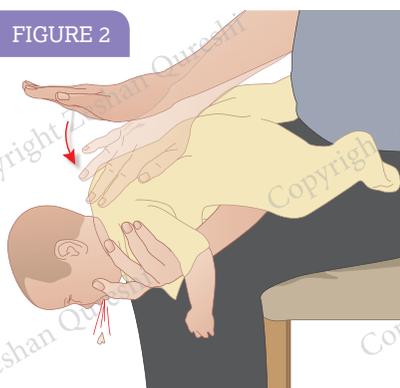
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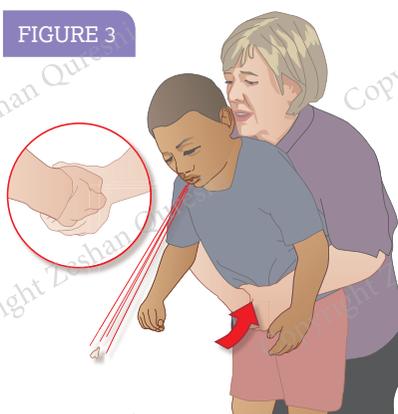
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FIGURE 2



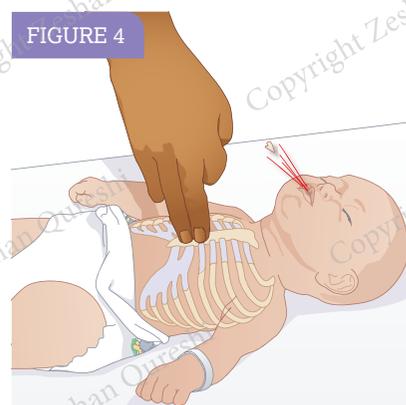
**Back blow.** The infant is placed over the leg to allow gravity to assist. The centre of the infant's back is struck firmly with the palm of the hand five times. In an older child, this is done keeping them upright, but bending them forward at the waist.

FIGURE 3



**Abdominal thrust.** Previously called the Heimlich manoeuvre. The person performing the manoeuvre makes a fist with one hand, and grabs it with the other. They then rapidly pull back and upwards through the abdomen to deliver each thrust.

FIGURE 4



**Chest thrusts.** This has a similar technique to chest compressions. Two fingers are placed at the base of the sternum. Five thrusts are then delivered, compressing downward about 1.5 inches.

(Figure 2) should be delivered, followed by five abdominal thrusts in children more than one-year-old (Figure 3), or five chest thrusts in children less than 1-year-old (Figure 4). The patient should be reassessed after each intervention, and the process should be repeated as necessary.

**Unconscious Child**

In a very severe obstruction, the child may become unconscious. In this case, attempt to remove anything seen in the mouth with one finger sweep, and then follow the paediatric basic life support, giving rescue breaths.

**INFECTION**

**Periorbital and Orbital Cellulitis**

**Periorbital Cellulitis**

Periorbital cellulitis is infection and inflammation of the skin and eyelid anterior to the orbital septum (unlike orbital cellulitis). This condition can be secondary to a sinus infection, but can also be due to a minor scratch or insect bite. In its most minor form, there is mild eyelid swelling without any systemic symptoms.

**Orbital Cellulitis**

Progress of periorbital cellulitis can lead to intra-orbital spread of infection

causing orbital cellulitis, with red flag signs of proptosis (bulging of the eye), limitation of eye movement and chemosis (conjunctival swelling). Colour vision and visual acuity are important to document, as these are sensitive features of optic nerve inflammation. A paramount concern is the potential for optic nerve injury and permanent loss of sight. Intracranial spread of the infection can occur on rare occasions.

**Management**

This condition is best managed with a multi-disciplinary approach involving paediatric, ENT, maxillofacial and ophthalmology teams.

Minor cases of periorbital cellulitis usually settle with oral antibiotics. Moderate cases need intravenous therapy. If concerns exist of intra-orbital infection, a computed tomography (CT) scan of the brain/orbit needs to be arranged urgently. Surgical intervention may require either an external or endoscopic approach to drain the pus, depending on the extent and position of the pathology.

**Tonsillitis**

**Aetiology**

Tonsillitis involves inflammation of the tonsils. It may be viral or bacterial in origin. Bacterial causes are more likely in the absence of cough, tender cervical

lymphadenopathy, high fever and tonsillar exudates. Classically, bacterial infection is with group A streptococcus.

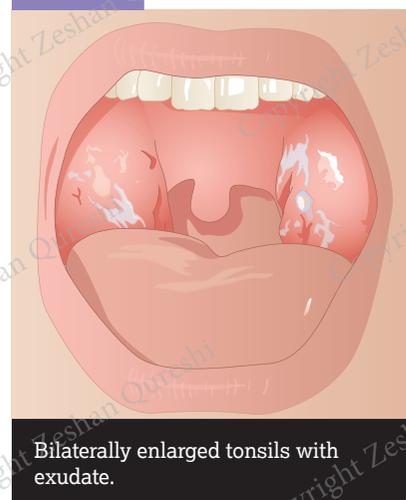
**Clinical Features**

Clinical features include:

- ▶ Sore throat.
- ▶ Pain on swallowing (odynophagia).
- ▶ Fever.
- ▶ Headache.
- ▶ Reduced oral intake.

On examination, the tonsils are red and enlarged with possible tonsillar exudate (Figure 5). Cervical lymph nodes may also be enlarged and tender. Simple analgesia, antipyretics and adequate

FIGURE 5



Bilaterally enlarged tonsils with exudate.





# 1.06

## ENDOCRINOLOGY

ZESHAN QURESHI, VAITSA TZIAFERI,  
SADHANANDHAM PUNNIYAKODI, MARYLYN-  
JANE EMEDO AND CLAIRE BRYANT

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### GLUCOSE REGULATION

#### Type 1 Diabetes Mellitus

Diabetes mellitus (DM) is a condition of elevated plasma glucose. There are two main types:

- **Type 1 diabetes (T1DM).** An absolute deficiency of the pancreatic hormone insulin.
- **Type 2 diabetes (T2DM).** Failure to appropriately make use of insulin to metabolise glucose.

The vast majority of cases in childhood are T1DM, although with the increasing obesity rates, T2DM is becoming more common. The World Health Organisation (WHO) has formulated a set of diagnostic criteria based on blood glucose measurements, dependent on the presence or absence of typical symptoms of the disorder (*Figure 1*).

#### Aetiology

T1DM is characterised by autoimmune T-cell mediated damage to the  $\beta$  islet cells of the pancreas. The  $\beta$  cells are responsible for insulin production and their destruction leads to insulin deficiency. Normally, insulin acts to reduce blood glucose through stimulating glucose uptake from the blood,

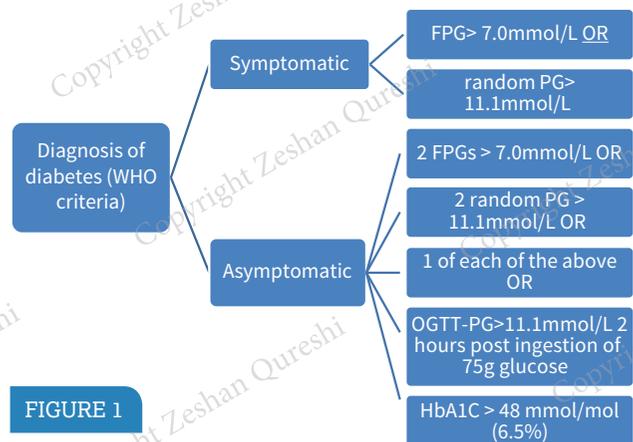


FIGURE 1

WHO criteria for diagnosis of Type 1 diabetes (FPG=Fasting Plasma Glucose, PG= Plasma Glucose, OGTT=Oral Glucose Tolerance Test). Adapted from "Definition and diagnosis of diabetes mellitus and intermediate hyperglycaemia": Report of a WHO/IDF consultation.



# 1.07

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MAXINE WILKE POOJA PAREKH, AND  
ZESHAN QURESHI

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# 1.08

## GENETICS

ISABEL MAWSON, CHRIS HARRIS, MAXINE WILKE AND ZESHAN QURESHI

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### APPROACHING GENETIC PROBLEMS

All cells in the human body, except gametes, have 46 chromosomes arranged into 23 pairs. Autosomes account for 22 of these pairs. The 23<sup>rd</sup> pair is made up of two sex chromosomes. In addition, every cell carries DNA within its mitochondria.

Through the processes of transcription and translation, genes enable each cell to build all the required proteins. Errors in an individual's genetic makeup can therefore affect every cell in the body. Genetic syndromes can be caused by problems at the gene or chromosome level or by errors in the packaging of the DNA. Detailed below are some of the mechanisms by which genes cause disease.

#### Patterns of Genetic Disease

##### Mendelian Inheritance

This describes a pattern of inheritance caused by mutations on a single gene, as seen in neurofibromatosis and Marfan syndrome.

##### Multifactorial, Polygenic Disorders

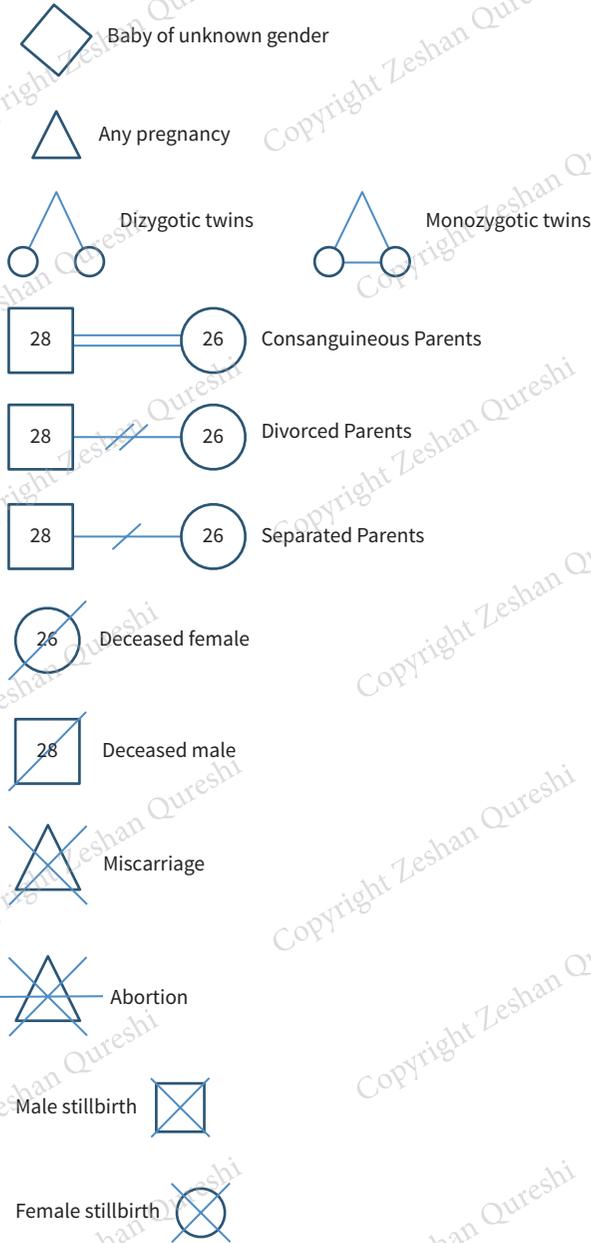
Some genetic mutations increase the risk of a disease, without necessarily causing the disease. For example, a mutation in the NOD2/CARD15 gene (which relates to the immune system) is associated with a slightly increased risk of Crohn's disease, though most people with the mutation remain healthy. This could be due to a certain environmental trigger required to make the genetic defect apparent, or it may be because other genes compensate for any problems related to the mutant gene. Such genes are said to have a low "penetrance", which means a low percentage of those with the defective gene (genotype) express the symptoms of the disease (phenotype).

##### Chromosomal Disorders

These disorders result from the absence or duplication of entire chromosomes. Examples include Down syndrome (Trisomy 21), Edward syndrome (Trisomy 18), Patau syndrome (Trisomy 13) and Turner syndrome (45,X).



FIGURE 5

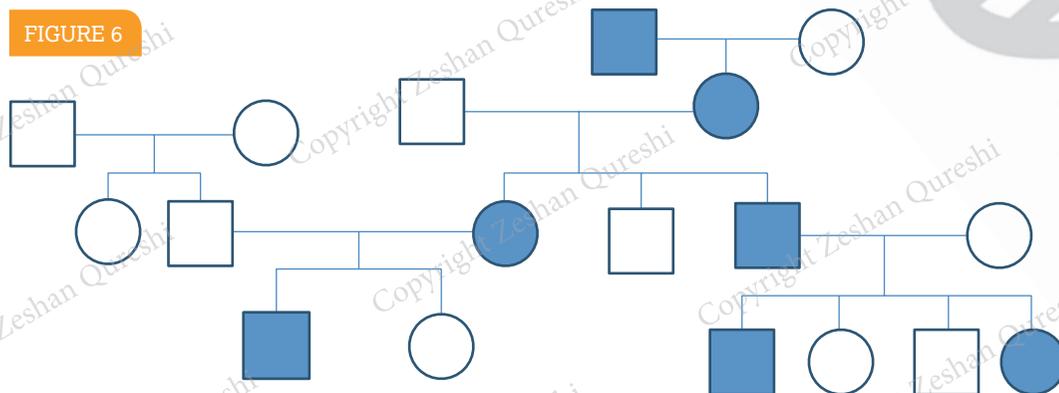


Other important symbols for pedigree diagrams.

TABLE 3: Autosomal dominant conditions

Condition	Gene/ Chromosome Affected	Characteristics
Achondroplasia.	FGFR3 gene (fibroblast growth factor receptor 3) on chromosome 4.	Short limbs, thoracolumbar kyphosis, disproportionately short stature, megaloccephaly with prominent forehead, midfacial hypoplasia.
Familial hypercholesterolaemia.	Three potential gene defects: LDLR (most common), APOB, PCSK9.	Premature coronary artery disease, elevated serum cholesterol levels.
Marfan syndrome.	Fibrillin 1 (FBN1) gene.	Heart defects, ophthalmological abnormalities, spontaneous pneumothorax, tall and thin, long limbs, spinal scoliosis and recurrent hernias.
Neurofibromatosis Type 1.	Caused by defects in the NF1 gene at 17q11.2 which produces neurofibromin.	6 or more café-au-lait spots >5mm in prepubertal and >15mm in postpubertal children. Neurofibromas, axillary freckling, Lisch nodules, optic glioma.
Neurofibromatosis Type 2.	Type 2 caused by defects in the NF2 gene at 22q11.2 which produces schwannomin.	Acoustic neuromas, meningiomas, neurofibromas, schwannomas, juvenile posterior capsular cataracts.

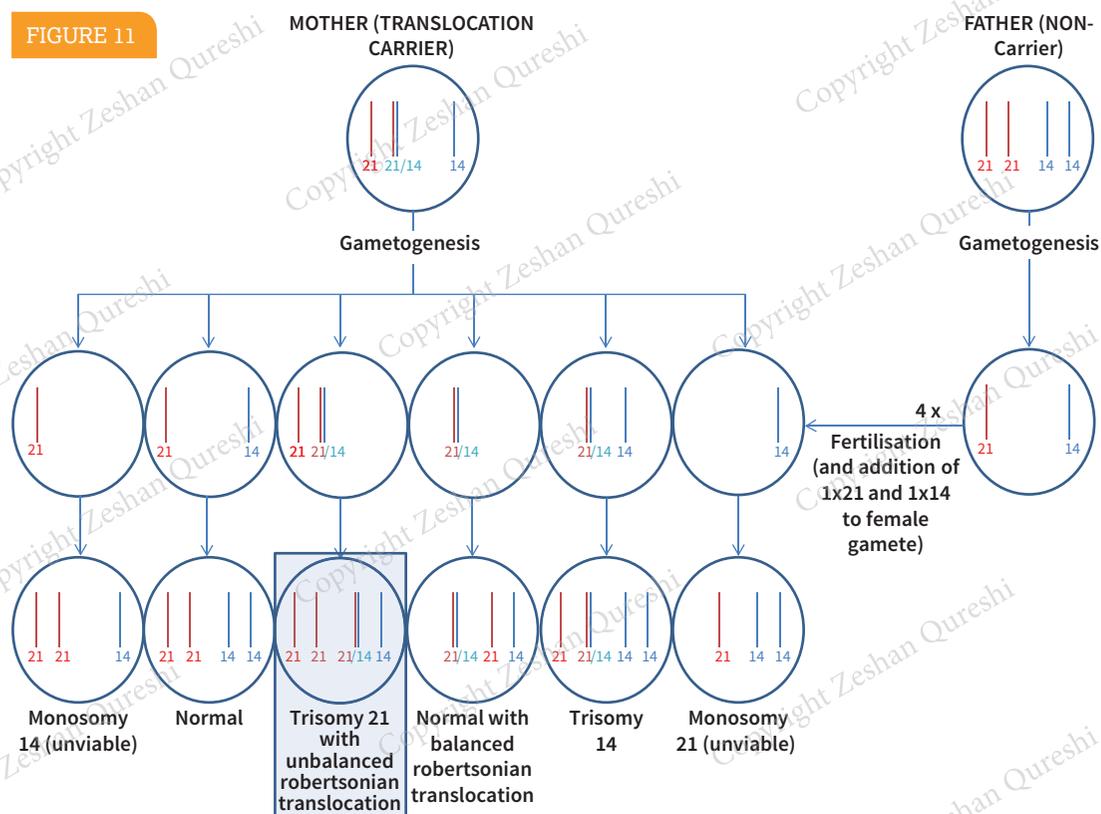
FIGURE 6



Pedigree diagram for an autosomal dominant disorder. All affected children must have an affected parent.

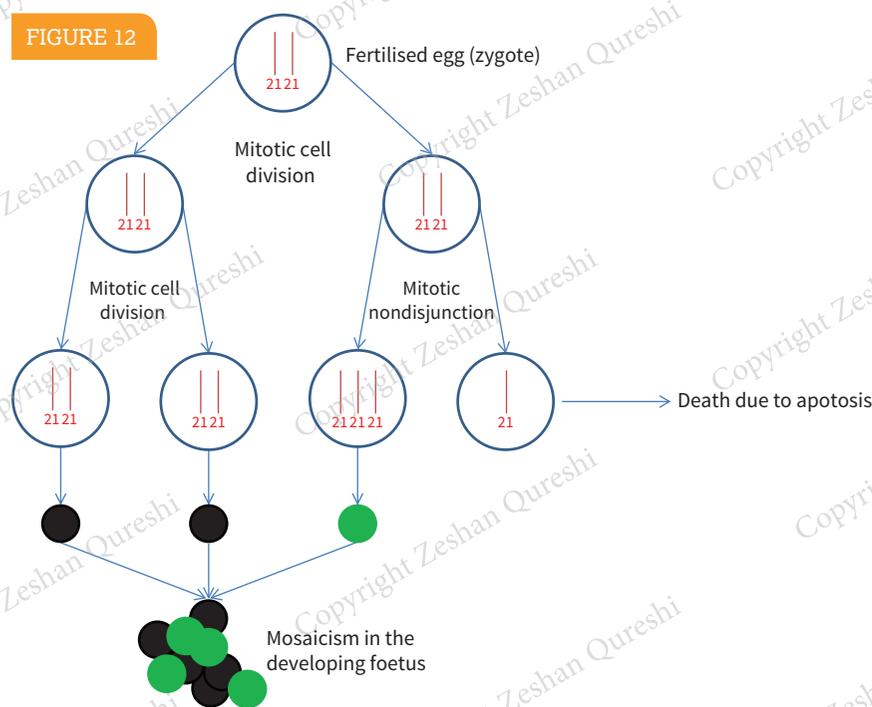


FIGURE 11



Robertsonian translocation, showing all six possible outcomes when a mother with a balanced Robertsonian translocation has a child with a father with normal chromosomes. The trisomy 21 cases are highlighted in blue.

FIGURE 12



Mosaicism caused by mitotic nondisjunction. The red line depicts one chromosome. Patients with mosaic Down syndrome usually have a less severe phenotype, although severity is very variable. Note that offspring will only be affected if gamete-producing cells carry the defect.





# 1.09

## HAEMATOLOGY

AMY MITCHELL AND ANNA CAPSOMIDIS

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### ANAEMIA

#### Overview of Anaemia

Anaemia is defined as a reduction in haemoglobin, and is due to a variety of possible causes. It is important to interpret the haemoglobin (Hb) level in relation to the age specific normal values (*Table 1*).

#### Aetiology

Three main processes lead to anaemia:

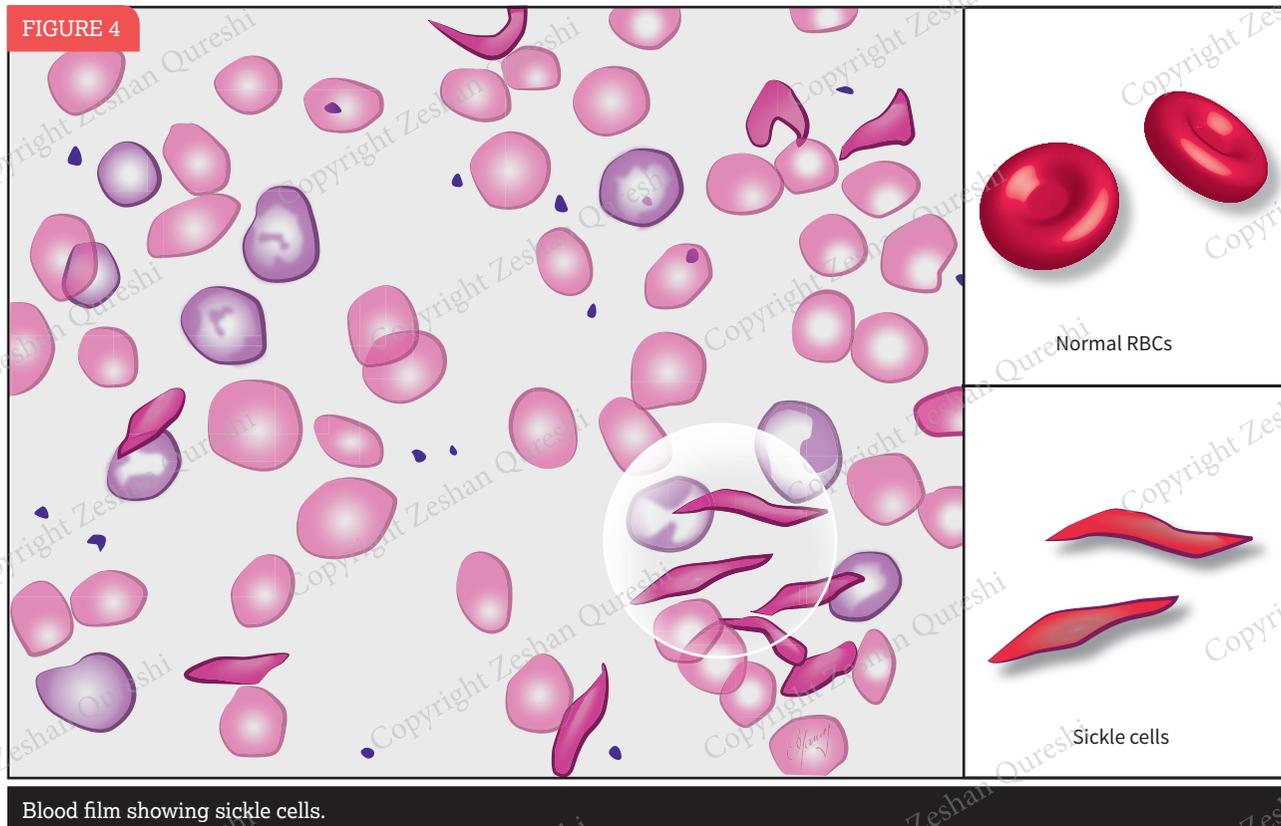
- 1 **Failure of red cell production.** e.g. iron deficiency, aplasia.
- 2 **Ineffective red cells.** e.g. haemoglobinopathies, sickle cell anaemia.
- 3 **Increased destruction of red cells.** e.g. hereditary spherocytosis.

Haemoglobinopathies are an important cause of anaemia. A haemoglobinopathy results in an abnormality in red cell structure. This may result in impaired oxygen carriage or decreased red cell life expectancy, thus giving rise to anaemia.

There are two categories of inherited haemoglobinopathies:

- 1 **Structurally abnormal haemoglobin molecules.** e.g. sickle cell anaemia.
- 2 **Impaired production of globin chains.** e.g. beta-thalassaemia.

The majority of haemoglobinopathies are autosomal recessive conditions. This means those with only one allele are asymptomatic carriers, i.e. sickle cell trait, beta-thalassaemia trait. Only homozygotes present with clinically detectable disease. These diseases have persisted in endemic areas because the asymptomatic presence of one allele (i.e. being a carrier) offers some protection from malaria.



### Management

Acute management is focused on identifying the underlying process and involves appropriate analgesia, oxygenation and treatment of associated sepsis, as outlined above.

Long-term outpatient management involves:

- ▶ Life-long prophylactic penicillin.
- ▶ Appropriate vaccinations (additional pneumococcal and influenza vaccines on top of routine schedule).
- ▶ Close monitoring for complications such as acute haemolysis, and painful episodes.

Corticosteroids, blood transfusions and hydroxycarbamide (which increases foetal Hb levels) may be helpful in those with severe episodes.

The requirement for long-term transfusions puts the child at risk of iron overload. This can be monitored by serum ferritin but more often now specialist MRI scanning techniques are being used to identify degree of iron overload. Other than limiting transfusions the other treatment of iron overload is to use an iron chelator such

as desferrioxamine usually via long-term subcutaneous infusion.

Haemopoietic stem cell transplantation (HSCT) is increasingly being utilised for haemoglobinopathies. The principle is replacement of the child's bone marrow, thereby fixing the underlying haemoglobinopathy. However, HSCT has important side effects, including risk of death during the transplant itself. HSCT is reserved for severe disease (four or more vaso-occlusive crises, acute chest crises despite therapy with hydroxycarbamine and disease in the central nervous system).

### Complications

Complications are shown in *Table 4*.

### Prognosis

Early identification of affected children in routine post-natal screening in many countries like the UK can be helpful in managing patients from an early age. In the UK, 99% of children with sickle cell survive to 16-years-old, but sickle cell accounts for 16% of all deaths in West African countries.

### Thalassaemia

Thalassaemia is a heterogeneous group of inherited conditions with autosomal recessive inheritance in which abnormal haemoglobin is produced in low quantity. As with sickle cell disease, the presence of thalassaemia trait (having one thalassaemia gene and one normal gene) confers some protection from malaria, thus giving carriers a selective survival advantage.

### Classification

Thalassaemias are classified according to which chain of the haemoglobin molecule is affected.

- ▶ In  $\alpha$ -thalassaemia, production of the  $\alpha$  globin chain is affected.
- ▶ In  $\beta$ -thalassaemia, production of the  $\beta$  globin chain is affected.

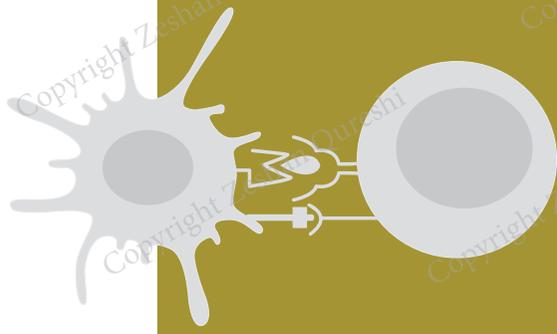
The most common clinical condition is  $\beta$ -thalassaemia major, which is characterised by a total absence of production of adult  $\beta$ -globin chains.  $\beta$ -thalassaemia intermedia is also autosomal recessive, but has milder symptoms.  $\beta$ -thalassaemia trait (with one normal gene) is usually asymptomatic.



# 1.10

# IMMUNOLOGY AND ALLERGY

MAANASA POLUBOTHU



## OVERVIEW OF THE IMMUNE SYSTEM

**T**he immune system combats the numerous microorganisms the body encounters daily. The body's immune response to infection has both innate and adaptive components. Disorders of the immune system can lead to immunodeficiency, allergy and autoimmune disease.

### Innate Immune System

The innate immune system refers to the immune defences that exist in all individuals, regardless of prior pathogen exposure. Therefore, innate immunity can be elicited immediately. It serves as the body's first-line defence. Its key components are detailed below.

#### Physical Barriers

- ▶ **Tight junctions.** These are found between cells in the skin and mucous membranes.
- ▶ **Secretions.** These include mucus, bile, enzymes and acid.

#### Cells that Secrete Inflammatory Mediators

- ▶ **Natural Killer Cells.** Cytotoxic lymphocyte cells able to identify foreign cells and trigger cell death.
- ▶ **Mast Cells.** White cells containing toxic granules including histamine which can be triggered to burst or "degranulate" upon activation.
- ▶ **Macrophages.** Large white blood cells capable of phagocytosis, able to engulf and digest cellular debris, microbes and foreign substances.

#### Cells that Detect Pathogens

- ▶ **Cells with Toll-like Receptors.** Receptors found on the membranes of macrophages and dendritic cells that detect microbes or parts of microbes (Pathogen-associated molecular patterns (PAMPS)).
- ▶ **Antigen Presenting Cells.** These cells are capable of engulfing and presenting foreign antigens to T cells to elicit an adaptive immune response. Examples include dendritic cells, some macrophages and some types of B cells.

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TABLE 3: Hypersensitivity reactions

Type	Onset	Mediation	Example
I.	Immediate in onset (usually occurs within 15-30 minutes of exposure).	This is mediated by IgE antibodies, which are produced in excessive amounts following sensitisation with the offending antigen (allergen). Upon exposure to the allergen and binding to IgE, IgE associated mast cells and basophils degranulate, releasing preformed mediators (e.g. histamine, tryptase, prostaglandins). Up to 20% of people will experience a late phase response, with symptoms recurring within the first 30 hours without repeat exposure to the allergen. The pathogenesis of this is unknown but is thought to be secondary to inflammatory changes initiated by the initial response (Figure 1).	<ul style="list-style-type: none"> <li>Asthma.</li> <li>Atopic conditions.</li> </ul>
II.	Delayed in onset (occurs within minutes to hours).	This is IgG mediated cell destruction. The complement system also has an important role. Antigens are usually endogenous, but may result from exogenous antigens attaching to the cell membrane (haptens).	<ul style="list-style-type: none"> <li>Goodpasture syndrome.</li> <li>Drug-induced haemolytic anaemia.</li> </ul>
III.	Delayed in onset (occurs within three to 10 hours of onset).	This is mediated by soluble immune complexes (mostly IgG but may be IgM). The complement system also has an important role. Antigens can be endogenous (e.g. SLE) or exogenous (e.g. chronic bacterial and parasitic infections).	<ul style="list-style-type: none"> <li>Rheumatoid arthritis.</li> <li>Systemic lupus erythematosus.</li> </ul>
IV.	Delayed in onset (peaking at 48-72 hours post exposure).	This is cell-mediated, via T-cells and macrophages.	<ul style="list-style-type: none"> <li>Contact dermatitis.</li> </ul>
V.	Delayed in onset.	This is endocrine receptor-mediated, resulting in impaired cell signalling.	<ul style="list-style-type: none"> <li>Graves' disease.</li> <li>Myasthenia gravis.</li> </ul>

ALLERGY

Hypersensitivity Reactions

An allergic reaction is an exaggerated reaction to an allergen in a normally functioning immune system. It comes under the broader remit of hypersensitivity reactions, which also includes autoimmunity. An allergen is a non-parasitic antigen that can elicit a type I hypersensitivity reaction.

Four types of hypersensitivity reactions are widely recognised. Type V is also included, though its existence is more debatable, as shown in Table 3.

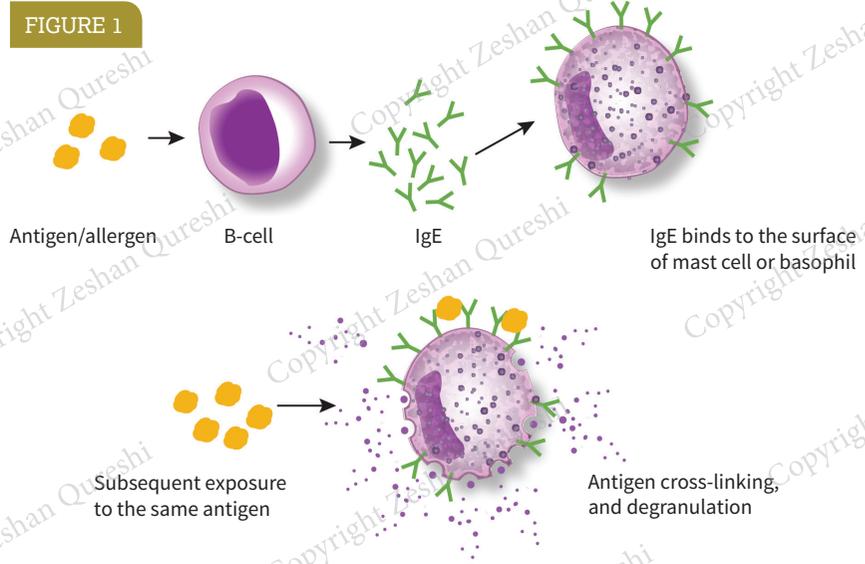
Atopic Triad

The atopic triad comprises:

- Eczema.
- Rhinitis.
- Asthma.

Classically, these develop in a typical sequence, alongside food allergy, and this is referred to as the allergic march (Figure 2). Atopic individuals are likely to have a genetic predisposition to Type I hypersensitivity reactions in response to common allergens. Circulating levels of IgE are elevated and immunoassays may reveal a positive response of IgE to a specific allergen. In such tests, the

FIGURE 1



Simplified scheme for IgE-mediated Type I hypersensitivity reaction. Mast cells/basophils are sensitised with allergen-specific IgE. On subsequent exposure, there is rapid degranulation, leading to the release of inflammatory cytokines and elicitation of a hypersensitivity reaction.

patient's serum is exposed to a specific antigen. If the patient has IgE antibodies that bind to the antigen, they are then visualised by the use of a second antibody that binds specifically to IgE.

Treatment of atopy is largely symptomatic and focuses on dampening the allergic response. Owing to its genetic component, atopy tends to run in families, although environmental

exposure to allergens plays a large part in the disease course. The prevalence of atopy is increasing in industrialised areas. The hygiene hypothesis postulates that this increased prevalence is the result of better hygiene, which has decreased human exposure to microbes. This has caused a shift in the immune response from the cells involved in fighting infection to those involved in

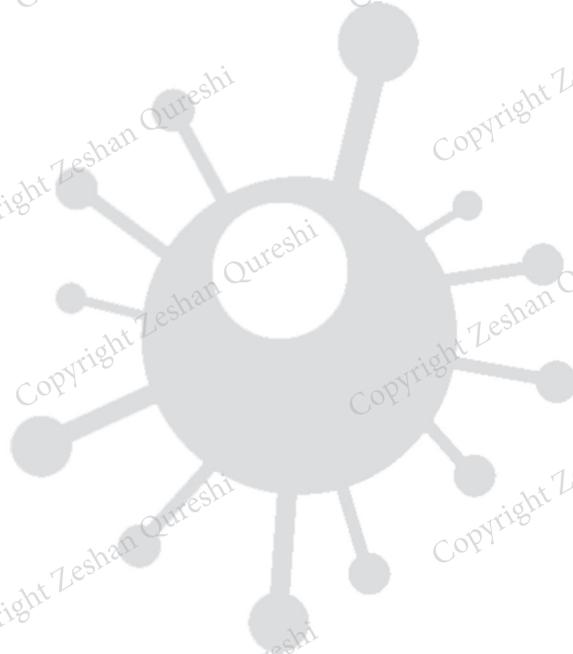




# 1.11

# INFECTION

PHILIPPA KING, ZESHAN QURESHI AND  
DAVID HO



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is supportive, with topical anaesthesia and oral rehydration salts if dehydrated. Rarely, children need to be admitted for intravenous fluids.

**Herpes Simplex Virus Type 2 (HSV-2)**

Genital herpes infection is typically caused by HSV-2 but can also be caused by HSV-1. Be wary of child sexual abuse presenting with genital herpes. The incubation period is around four days, and again prodromal symptoms include itching and pain prior to the onset of genital lesions. Other symptoms include headache, fever, myalgia and back pain. Other manifestations of herpes include meningoencephalitis, eczema herpeticum and eye infections, for which intravenous antiviral agents may be required.

**Maternal Genital Herpes**

This can potentially cause congenital herpes infection following intrauterine exposure or vertical transmission after vaginal delivery. Neonatal herpes infection can result in central nervous system infection (meningoencephalitis) and disseminated infection with significant risk of disability and death, even if treated (IV aciclovir).

**Human Herpes Virus Type 6 (HHV-6)**

This is the most common cause of roseola infantum, with an incubation period of about 10 days. In this condition, a child 6-months-old to 2-years-old presents with two to three days of fever, but then as the fever subsides, a widespread rash emerges. It is also known as “sixth disease”, as the rash often emerges on day 6 of the illness. Often, parents get anxious as the child appeared to have been getting better (due to fever subsiding) before the rash emerged, but the condition is benign, requiring no treatment. HHV-6 is also implicated in encephalitis, hepatitis and febrile convulsions.

**Varicella Zoster Virus**

Varicella-zoster virus (VZV), also known as chickenpox, is a common childhood

infection (Figure 1). Children acquire VZV through contact with infected individuals at nurseries, schools or social groups.

The incubation period is 10 to 21 days. Prodromal features include fever, malaise, headache and abdominal pain, usually one to two days before the onset of the rash. The itchy rash is characteristic and begins as macules, progressing to papules and then vesicles which can crust over. These lesions can turn haemorrhagic. The child will be infectious from 48 hours before the onset of the rash until all the lesions have crusted over.

The diagnosis is commonly made clinically, although swabs can be taken and PCR performed if required.

Treatment with antiviral medication is unnecessary in healthy children, as most VZV infections are self-limiting. However, treatment is indicated in asymptomatic neonates (if the mother acquires chickenpox within seven days prior to delivery, or up to four days afterwards), in adolescent children and in immunocompromised children who tend to have more severe disease. Varicella-zoster immunoglobulins are used in certain high risk groups.

If fever persists in children with VZV, complications such as bacterial infections caused by *Staphylococcus aureus* and *Streptococcus pyogenes* need to be considered and treated. Other important complications include cerebellar ataxia, varicella encephalitis, pneumonitis, hepatitis, thrombocytopenia and nephritis.

**Shingles**

Reactivation of VZV typically occurs in adulthood, causing painful eruptions of vesicles in a dermatomal distribution (Figure 2). This is known as herpes zoster or shingles. Antiviral medication such as aciclovir and valaciclovir can shorten the duration of pain and lead to quicker resolution of lesions. A shingles vaccine is also now available and routinely offered to elderly adults.



FIGURE 1

Chicken pox (D@nderm).

FIGURE 2



Varicella zoster infection of the V division of the trigeminal nerve.





# 1.12

## INTENSIVE CARE

KUNAL BABLA AND SAM THENABADU



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### INTRODUCTION

Intensive care is suitable for patients with potentially recoverable life-threatening conditions who would benefit from more detailed observation, treatment and technological support than is available in general wards or high dependency facilities. Complex monitoring equipment is available (*Table 1*), as well as more advanced interventions. It is also recognised that end-of-life management, including potential organ donation and family bereavement care are integral to caring for critically ill children.

The paediatric intensive care unit serves the most unwell children, who present with a range of both common and rare clinical conditions. The caseload varies throughout the year but will include a mix of emergency medical, surgical and trauma cases from both within the designated paediatrics hospital and the paediatric transfer team from the regional hospitals it supports. Certain planned surgeries may require a high level of airway support or monitoring post-operatively and may also command the need for ICU beds.

The clinician's priorities, at the bare minimum, involve:

- ▶ Regimented airway management and ventilator support.
- ▶ Cardiovascular monitoring, invasive circulatory support, with renal assessment and close fluid management.

In addition, assessment of the child's nutrition, microbiology, radiology, and invasive lines and resuscitation status are all carefully considered on a daily basis.

It is important for the paediatric intensive care unit (PICU) team to remember that as well as delivering high-quality medical care, the team must involve the child's family in the care plan. This places heavy emphasis on clear and effective communication on all aspects of their child's care. This can often involve difficult conversations about how unwell a child is or even when it might be appropriate to consider palliative care. The PICU clinician should be sensitive to a family's wishes and views on their child's care.



# 1.13

## CHILDREN AND THE LAW

ZESHAN QURESHI



### INTRODUCTION

Children are a vulnerable group and may not be able to represent their own wishes. Governments therefore place significant emphasis on making sure children's rights are protected and that their needs are adequately represented. This section will predominantly reflect law in England and Wales, but the underlying principles are more universal.

Children's rights are enshrined in the United Nations Convention on the Rights of the Child, which states:

- ▶ Actions taken concerning a child must have their best interests as a primary consideration.
- ▶ If the child can voice their opinion, they should be able to express it freely.
- ▶ The child's view should be given due weight (based on age and maturity).

In the UK, the Children Act 2004 states that the welfare of the child is paramount, and all professionals dealing with children have the duty to put the child at the centre of their actions. Similar legislation exists across the world.

### Incorporating Multiple Viewpoints

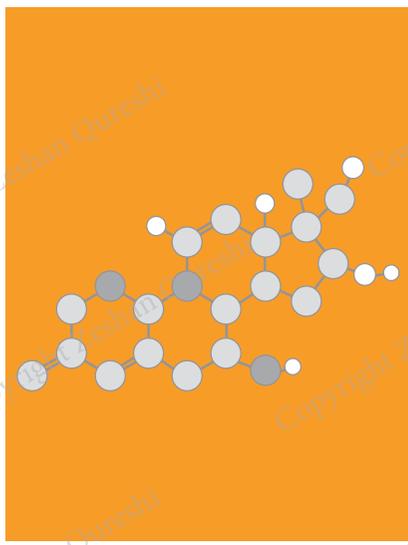
In making decisions for children, one should try to incorporate the views of:

- ▶ **The child.** This includes both previously expressed beliefs and current wishes. Someone under 16-years-old may be deemed to have capacity, but even if they do not, their wishes and beliefs must be taken into account, as far as it is in their interests to do so.
- ▶ **The child's parents.** They may provide valuable insight into their child's wishes, beliefs and normal behaviour. Looking at the cultural background of the child and speaking to friends and other relatives may help as well in this regard.
- ▶ **The multidisciplinary team.** This will include nurses, physiotherapists, school teachers and any other professionals involved in the child's care.

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# 1.14

## METABOLIC MEDICINE

ZESHAN QURESHI, STEPHANIE CONNAIRE  
AND ZAINAB KAZMI

### WHAT IS METABOLIC DISEASE?

**M**etabolic disease results from defects in enzymes involved in metabolic pathways of the body leading to accumulation of toxic metabolites, deficiency of essential metabolites or both.

Principles of management therefore focus on reversing catabolism, replacing the deficiency and chelating or excreting toxic metabolites. Symptoms and signs as well as clinical course relate to which pathway is involved.

The term “inborn error of metabolism” refers to metabolic disorders that are inherited, usually as a result of a single gene defect affecting a significant enzyme in a metabolic pathway. Hundreds of different metabolic disorders have been described. Although each metabolic disorder is individually rare, collectively, they are not uncommon, with around 1 in every 2500 children in the UK born with an inherited metabolic disease.

Metabolic medicine is becoming increasingly important, particularly in the context of newborn screening, where these diseases can be detected at the pre-symptomatic stage. In the UK today, the diagnosis of medium-chain acyl-CoA dehydrogenase deficiency (MCADD) is given via a phone call to the parents of a well baby with a positive screening test, rather than when the child presents with hypoglycaemia and its complications.

This chapter will give a brief introduction regarding when to suspect metabolic disease and an overview of a few common forms of these diseases.

### WHEN SHOULD METABOLIC DISEASE BE SUSPECTED?

The wide variation and non-specific presentation of metabolic disease means that a high index of suspicion is required to make the diagnosis. Affected children who are not diagnosed through neonatal screening may present as acutely unwell, with an undiagnosed metabolic disorder. Metabolic disorders tend to present at times of metabolic stress, for example, during the neonatal period (when presentation can be easily mistaken as sepsis), at times of dietary change or with intercurrent illness. Clues can be found in the history, examination and investigations.

### Newborn Screening

The neonatal heel prick or Guthrie test is a screening test performed on newborn babies at day five to day eight of life.

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COMMON METABOLIC DISORDERS

Galactosaemia

Aetiology

Lactase breaks down dietary lactose to glucose and galactose. Galactose can then be mobilised for energy, via the pathway described in *Figure 1*. Galactosaemia can result from deficiency in any of the three enzymes: galactokinase, galactose-1-phosphate uridyl transferase (GALT) and UDP galactose-4-epimerase. Enzyme defects lead to an elevated blood galactose level. The most common and severe form is from GALT deficiency.

Clinical Features

Affected infants will often present in the neonatal period with jaundice, hepatomegaly, coagulopathy and cataracts. Older children may present with faltering growth or renal pathology. Galactosaemia is associated with *E.coli* sepsis.

Investigations

Diagnosis is made by testing urinary reducing substances and a Gal-1-PUT (galactose-1-phosphate uridyltransferase) assay. Liver function tests may be deranged, with prolonged jaundice and possible coagulopathy. Affected children should have an eye examination.

Management

Management of galactosaemia is through a lactose/galactose free diet (soya based formula and then dairy-free diet) and ensuring adequate calcium and Vitamin D intake.

Prognosis

The immediate prognosis is good with early diagnosis and appropriate treatment. Death from liver failure or sepsis is possible if appropriate dietary changes are not made.

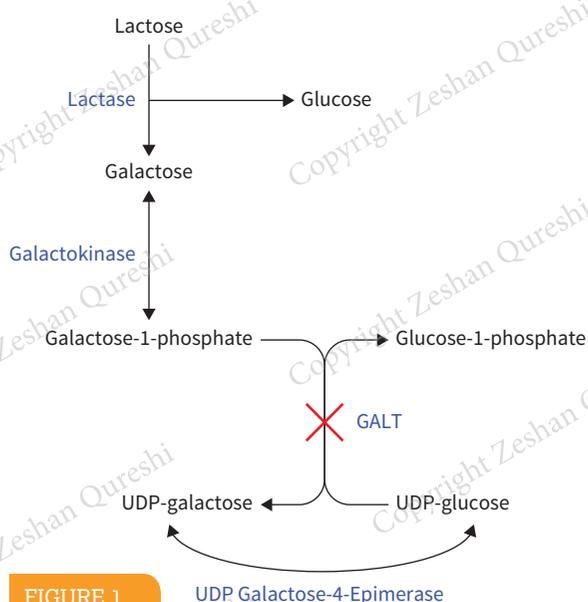


FIGURE 1 UDP Galactose-4-Epimerase  
Galactose metabolism and GALT deficiency.

Hypogonadotrophic hypogonadism is common in women, with a high risk of ovarian failure. There may also be developmental delay.

Glycogen Storage Disease (GSDs)

This section will mainly focus on GSD type 1, with a general overview of other types.

Aetiology

Glucose is stored as glycogen in the liver and muscles, with specific enzymes involved in glycogenolysis (*Figure 2*). Individuals affected by GSDs have a defect in one of these enzymes, leading to glycogen build-up (and intermediate compounds), plus difficulty maintaining normoglycaemia. GSDs are categorised according to the enzyme defect involved (*Table 3*). Type 1 GSD is the most common.

Clinical Features

Some disorders affect the liver, some affect the muscles and some affect both (*Table 3*). The main role of glycogen in the liver is to maintain glucose homeostasis; therefore, GSDs affecting the liver may lead to hypoglycaemia and hepatomegaly. GSDs affecting the muscles cause weakness and fatigue.

Investigations

Investigations include checking glucose, lactate, uric acid and lipids. Confirmation of the diagnosis is via enzymology assays or genotyping, depending on the subtype. Liver biopsy may be needed in some cases.

Management

For most types of GSD, no specific treatment is available and therefore symptomatic management is important. Hypoglycaemia may occur frequently, particularly during periods of starvation. In the long term, liver transplantation may be necessary.

In GSD 1a, regular feeding during the day is essential, including continuous feeding overnight, with slow release carbohydrates (e.g. uncooked corn starch in those over two) to facilitate longer gaps between feeds. As there is a risk of gout from hyperuricemia, allopurinol should be given. GSD 1b requires the addition of septrin or granulocyte colony-stimulating factor in view of neutrophil dysfunction.

Prognosis

The prognosis for GSD type 1 depends on glucose control. Complications include short stature, renal disease, osteoporosis and liver adenomas (which may become malignant).

Homocystinuria

Aetiology

Homocysteine is an amino acid formed by converting methionine to cysteine. Homocystinuria is a rare inherited metabolic disorder characterised by high concentrations of





# 1.15

## NEONATOLOGY

CHRISTOPHER HARRIS AND  
ZESHAN QURESHI



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**FIGURE 11**  
Mongolian blue spot.



**FIGURE 12**  
Port wine stain.

although lesions found on the nape of the neck may persist into adulthood.

**Port Wine Stain**

Port wine stains (also known as naevus flammeus) result from low-flow vascular malformations of the dermal capillaries and post-capillary venules (Figure 12). These flat, blanching, pink-red lesions are present from birth. They can be found anywhere on the body, but tend to be unilateral with midline demarcation. These lesions do not regress and may become darker and thicker with age. Complications include thickening, nodularity and bleeding. The majority of these lesions occur as isolated lesions but they may occur as part of a syndrome (e.g. Sturge-Weber syndrome: facial port wine stain, ipsilateral venous malformation in the meninges and ocular abnormalities). Glaucoma is associated with periocular port wine stains; therefore, neonates with port wine stains in the distribution of the ophthalmic or maxillary branches of the trigeminal nerve should be referred for ophthalmology review. The presence of a facial port wine stain and glaucoma or neurological symptoms should rouse suspicion of Sturge-Weber syndrome and a neurological review and brain imaging should be sought. Treatment aims are improvement of the aesthetic appearance and prevention of complications. This may involve repeated treatments with pulsed dye laser therapy.

**Benign Pustular Melanosis**

This consists of superficial pustules, present at birth, which rupture easily without any actual pus content, leaving a spot of hyperpigmentation (Figure 13). Pustules usually last for one or two days, although the hyperpigmentation may persist.

**Melanocytic Naevi**

Melanocytic naevi represent benign proliferations of a subtype of melanocyte cell known as the naevus cell (Figure 14). They can be classified as congenital (occurring from birth) or as acquired. Congenital melanocytic naevi tend to grow rapidly in infancy, starting as flat lesions with even pigment,



**FIGURE 13**  
Benign pustular melanosis.



A

**FIGURE 14**

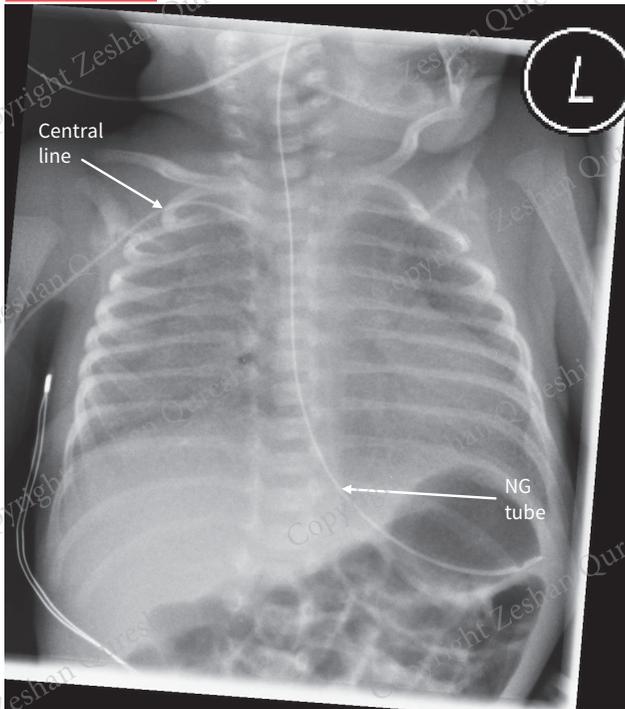


B

A) Small naevus on the face B) Giant naevus.



FIGURE 33



Respiratory Distress Syndrome. Note the interstitial shadowing bilaterally, giving a ground-glass appearance. An NG tube is present in the stomach and a central line is present, entering from the right arm.

**Aetiology**

Those most likely to be affected are extremely premature neonates. Other risk factors include infection and postnatal lung injury from ventilator support.

**X-Ray Appearance**

The appearance on X-ray begins with that of RDS (Figure 34), but then can evolve into that of pulmonary interstitial emphysema (Figure 35).

**Management**

Management involves nutrition optimisation, oxygen supplementation and ventilation support as required. Corticosteroid therapy and/or diuretic therapy may be helpful in improving lung function. Many babies with chronic lung disease will get better as they grow; some go home on oxygen and perhaps remain ventilator-dependent or die subsequently of the disease.

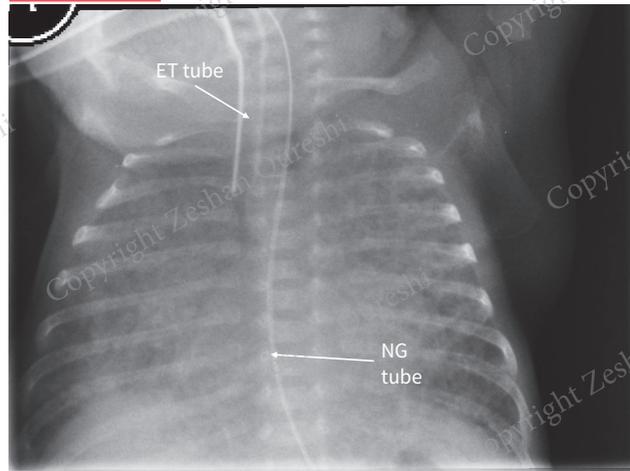
**Transient Tachypnoea of the Newborn**

This is the most common cause of respiratory distress in term babies, and is due to a delay in clearance of foetal lung fluid.

**Aetiology**

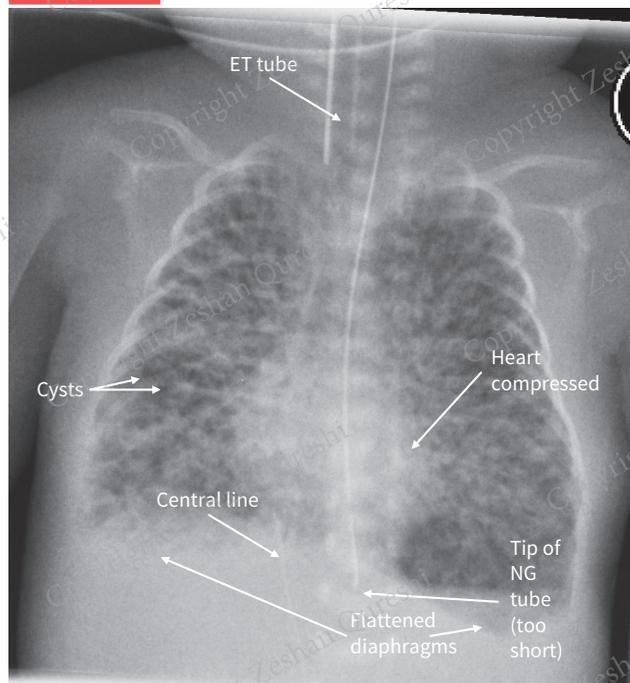
It is more common following delivery by caesarean section. The exact pathophysiology is unclear, but it is likely due to a lack of exposure of the infant to hormones and stresses involved during labour.

FIGURE 34



Diffuse interstitial shadowing in evolving bronchopulmonary dysplasia. Note also there is an NG tube in situ and an endotracheal (ET) tube sitting at the level of T3.

FIGURE 35



Broncho-pulmonary dysplasia. There is diffuse interstitial shadowing throughout the lung fields. There is hyperexpansion of the lungs, due to air trapping, resulting in flattening of the diaphragms. Cysts are visible in both lung fields. The heart also appears smaller, as the elevated intrathoracic pressure reduces venous return. Note also the ET tube present at the level of T1 and the central line at approximately T11. The nasogastric tube is short, only just crossing the diaphragm.

**X-Ray Appearance**

Radiological features may include cardiomegaly, pleural effusions, fluid in the horizontal fissure and prominent perihilar interstitial markings (Figure 36).





# 1.16

## NEUROLOGY

JOHN JUNGPA PARK, ZESHAN QURESHI  
AND DEBASREE DAS



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**Spastic Cerebral Palsy**

Spastic cerebral palsy results from upper motor neurone injury and thus manifests with classic signs of upper motor neuron injury, including:

- ▶ Muscle weakness (particularly antigravity muscles e.g. dorsiflexion of the feet).
- ▶ Increased muscle tone.
- ▶ Brisk tendon reflex.
- ▶ Sustained clonus.

Depending on the location of the insult different parts of the body are affected. Three subtypes of spastic CP are recognised: spastic hemiplegia,

spastic diplegia and spastic quadriplegia (**Table 7**).

**Extrapyramidal Cerebral Palsy**

Extrapyramidal cerebral palsy (also known as athetoid, choreoathetoid or dyskinetic cerebral palsy) arises in response to damage to the extrapyramidal tracts in the brain (particularly the basal ganglia).

Therefore, upper motor neurone signs are absent (**Figure 4**). Causes include hypoxic-ischaemic injury during delivery and kernicterus.

Infants are initially hypotonic, with poor head control, but show

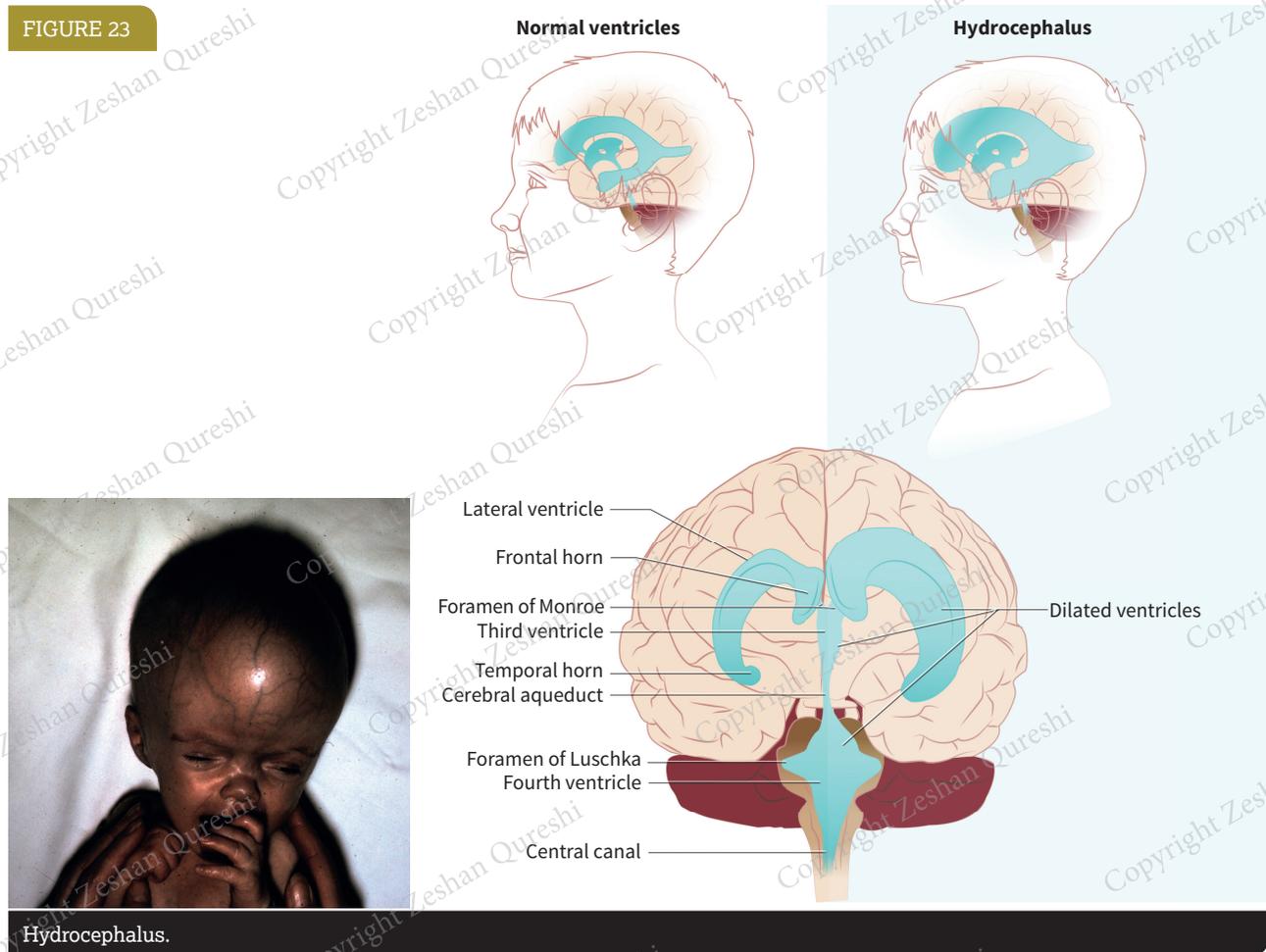
increased tone and dyskinetic movements as they develop. Dyskinetic movements include:

- ▶ **Dystonia.** A broad term describing involuntary, sustained contractions of opposing muscle groups. This leads to abnormal posture, twisting movements and repetitive movements.
- ▶ **Athetosis.** Slow, involuntary, writhing movements.
- ▶ **Chorea.** Brief, irregular movements that are non-repetitive/rhythmic. They appear to flow smoothly, giving them the appearance of dance-like movements.

**TABLE 7: 3 Subtypes of Spastic Cerebral Palsy**

	Affected body parts	Presentation	Additional Features
 <p>Spastic Hemiplegia.</p>	The arm and leg on one side, with arms affected more, and sparing of the face. Spasticity greatest in antigravity muscles.	<p><b>Affected arm:</b></p> <ul style="list-style-type: none"> <li>• Flexed, pronated arm, with fisting of the hand.</li> <li>• Hand preference before 12 months.</li> <li>• Difficulty in hand manipulation.</li> </ul> <p><b>Affected Leg:</b></p> <ul style="list-style-type: none"> <li>• Circumduction gait on affected side (stiff leg, not flexed at knee/ankle, rotated away and then towards the body with each step).</li> <li>• Toe-heel (tip toe) gait on affected side.</li> <li>• Delayed walking.</li> </ul>	<ul style="list-style-type: none"> <li>• May have growth arrest in the extremities (particularly if parietal lobe affected).</li> <li>• Moderate incidence of seizure disorders, and intellectual impairment.</li> </ul>
 <p>Spastic Diplegia.</p>	Lower limbs (with some involvement of upper limbs). Referred to as paraplegia if no involvement of upper limbs.	<ul style="list-style-type: none"> <li>• Commando crawl (dragging legs while crawling).</li> <li>• Scissoring of the lower legs.</li> <li>• Severely delayed walking.</li> <li>• Toe-heel (tip toe) gait, with feet held in equinovarus position.</li> <li>• Legs may be significantly underdeveloped compared to the arms.</li> </ul>	<ul style="list-style-type: none"> <li>• May have disuse atrophy, and impaired growth of the lower extremities compared to upper extremities.</li> <li>• Usually have normal intellectual development. May have learning difficulties and deficits in other neurological pathways (e.g. hearing).</li> <li>• Minimal risk of seizures.</li> </ul>

FIGURE 23



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# 1.17

## NUTRITION

CHI HAU TAN



### FEEDING

#### Breastfeeding

Breastfeeding has many advantages and breast milk is considered the ideal food for infants (*Table 1*). The World Health Organization (WHO) has recommended exclusive breastfeeding (i.e. no other fluids or solids) until an infant is 6-months-old. Breastfeeding is encouraged until the baby is at least 12-months-old, as long as both the mother and child desire it. *Box 1* lists some contraindications.

#### Jaundice and Breastfeeding

Breastfed babies are more likely to become jaundiced in the neonatal period. This may be for two reasons:

- ▶ If breastfeeding is slow to establish, a baby may lose significant weight in the first week of life. Dehydration due to insufficient intake exacerbates jaundice.
- ▶ Breast milk innately inhibits certain liver enzymes involved in bilirubin excretion (glucuronyl transferase), making jaundice more prominent.

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TABLE 1: Benefits of breastfeeding

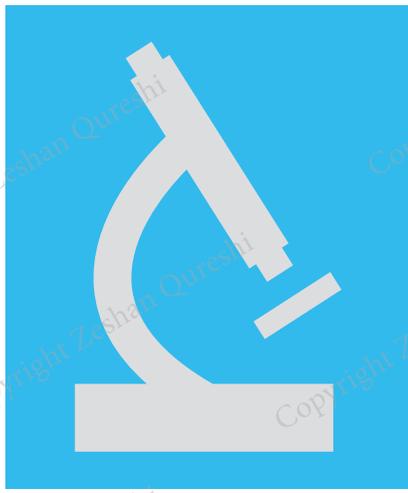
#### For infants

- Provides the right composition of nutrients for healthy growth and development.
- Readily absorbed by the body.
- Not complicated by infections associated with formula milk preparation.
- Contains antibodies.
- Numerous long-term health benefits, including improved visual acuity and cognitive development, and reduced risk of developing allergies, inflammatory bowel disease, cardiovascular diseases, diabetes and obesity.

#### For mothers

- Stimulates release of oxytocin that helps with uterine contraction and reduces postpartum haemorrhage.
- Facilitates postpartum weight loss.
- Contraceptive effect through prolongation of lactational amenorrhoea (if used exclusively).
- Lowers the risk of breast cancer, ovarian cancer and osteoporosis.
- Saves the time and expenses associated with preparation of formula milk.
- Provides opportunity for infant-mother bonding.





# 1.18

## ONCOLOGY

ANNA CAPSOMIDIS AND AMY MITCHELL

### PRINCIPLES OF ONCOLOGICAL CARE

#### Presentation

Leukaemia, lymphoma and brain tumours are the most common cancers affecting children. As childhood malignancy is rare, there can be a delay in diagnosis after presenting symptoms first emerge. Timely diagnosis and specialist referral can improve outcome. Children referred to paediatric oncology centres for suspected cancer require many investigations to make a histological diagnosis and to assess for metastatic spread before treatment starts.

Management of childhood malignancy depends on:

- ▶ Cancer type.
- ▶ Metastasis.
- ▶ Child's age.
- ▶ Co-morbidities.

Whilst the mainstay of treatment for haematological malignancies is chemotherapy, treatment of solid tumours usually requires a combination of chemotherapy, surgery and radiotherapy.

#### History

- ▶ Persistent unexplained fever, lethargy or weight loss: after excluding common infections (such as upper respiratory tract infection [URTI], urinary tract infection [UTI] or pneumonia), consider malignancy, atypical infections, inflammatory bowel disease (IBD), and human immunodeficiency virus (HIV).
- ▶ Unexplained bleeding from mouth, gums or nose.
- ▶ Unexplained bruises.
- ▶ Persistent bone pain or back pain (including nocturnal, which may wake the child from sleep).
- ▶ New onset limp, or a child who has stopped weight bearing.
- ▶ New lump or mass.
- ▶ Headaches for more than two weeks severe enough to wake the child from sleep in the morning, or associated nausea and vomiting.
- ▶ Shortness of breath (especially when associated with suspicious lymphadenopathy).
- ▶ Unexplained new onset seizures.
- ▶ Persistent, unexplained vomiting.
- ▶ Parental anxiety with no clear diagnosis.
- ▶ Behavioral change.
- ▶ Visual loss, new onset squint (can be paralytic e.g. due to a 6th nerve palsy, or non-paralytic e.g. brainstem lesion).
- ▶ Rapid head growth in under 2-year-olds.

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can be delivered in the child's own home, in hospital, or in a children's hospice. Traditionally, children were referred to the paediatric palliative care team when their disease was no longer curable. However, more recently a "parallel planning" approach has become more popular. The aim of this approach is for much earlier involvement and collaborative working to enable appropriate symptomatic management alongside curative treatment.

Making end-of-life decisions is extremely challenging for children, their families and the healthcare professionals involved. Communication is essential to respect the best interests of the child. Children and young people should be involved in end-of-life discussions as much as possible, unless this is deemed harmful to the child.

**COMMON MALIGNANCIES**

**Leukaemia**

Leukaemia is the most common childhood cancer, with approximately 400 new cases in the UK each year.

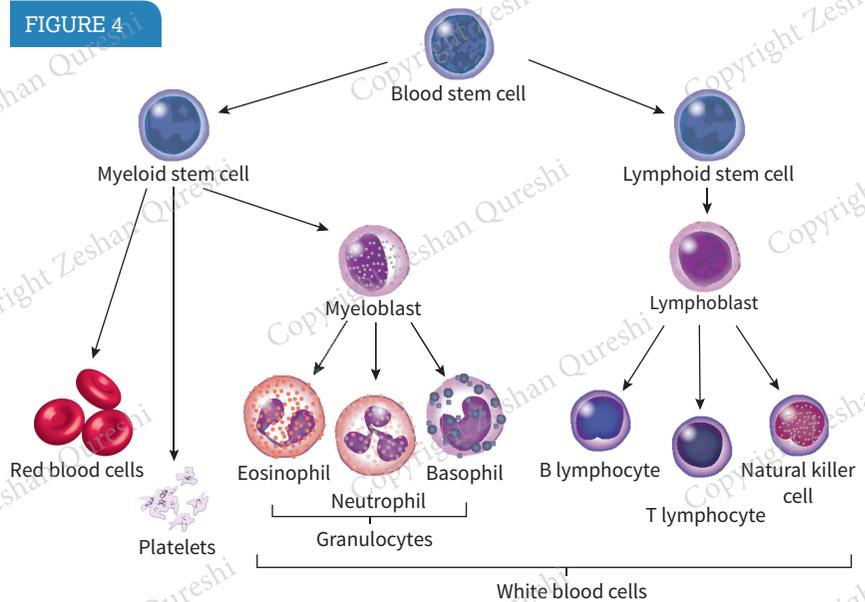
**Aetiology**

Leukaemia is a haematological malignancy caused by abnormal cellular division of immature blood cells (lymphoblasts or myeloblasts) in the bone marrow. Clinical presentation reflects the infiltration of leukaemic cells into normal tissues including the bone marrow, lymph nodes, liver, spleen, brain and testes:

- ▶ Bone marrow failure leads to anaemia (pallor), thrombocytopenia (bleeding, bruising and petechiae) and neutropenia (infections).
- ▶ Tissue infiltration leads to features such as bone pain, hepatosplenomegaly and lymphadenopathy.

Leukaemia is classified into acute or chronic, as well as into myeloid or lymphoid, according to the cell of origin (Figure 4). Various genetic and inherited factors have been shown to play a role in the development of leukaemia (Box 1).

FIGURE 4



Normal haematopoiesis showing the lymphoid and myeloid cell lines.

Siblings of those affected also have a higher risk of developing the disease.

Approximately 80% of childhood leukaemias are acute lymphoblastic leukaemias (ALL) and 15% acute myeloid leukaemias (AML). Key differences between ALL and AML

are summarised in Table 2, but they generally present with similar symptoms. Chronic lymphocytic leukaemia (CLL) and chronic myeloid leukaemia (CML), although common in adults, are extremely rare in children.

**Box 1: Genetic Syndromes associated with increased risk of childhood leukaemia**

- Down Syndrome.
- Fanconi anaemia.
- Ataxia telangiectasia.
- Neurofibromatosis type 1.
- Wiskott-Aldrich syndrome.
- Noonan syndrome.

**Clinical Features**

Children with ALL usually present with a short history over days or weeks, whereas the symptoms and signs of AML may develop more slowly. Initially, symptoms may appear very non-specific and mimic a viral infection.

Common presenting symptoms and signs include:

- ▶ Fever.
- ▶ Pallor.
- ▶ Petechiae and bruising.
- ▶ Tiredness or lethargy.

TABLE 2: Acute lymphoblastic leukaemia vs. Acute myeloid leukaemia

	ALL	AML
Cell line affected.	Lymphoid.	Myeloid.
Percentage of leukaemia in children.	80%.	15%.
Peak age incidence.	2 to 3-years-old.	Under 2-years-old.
Sex predominance.	Boys > Girls.	Boys = Girls.
Auer rods.	Auer rods not present.	Auer rods usually present.
Prognosis (children <14-years-old).	Approximately 90% five-year survival rate.	Approximately 60-70% five-year survival rate.





# 1.19

## ORTHOPAEDIC AND RHEUMATOLOGICAL DISORDERS

ANAND GOOMANY AND ALEXANDER YOUNG

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► **Displaced fractures.** Urgent reduction is required. Closed reduction techniques followed by splinting can be used for type I and II fractures. Type III and IV fractures may be amenable to closed reduction but will require open reduction and internal fixation if accurate reduction is not achieved.

**Prognosis**

These fractures must be carefully reduced to minimise the risk of premature ossification of the affected part, causing serious growth arrest and deformity. Salter-Harris type I and II fractures rarely result in growth arrest or deformity, in contrast to types III-V where the risk of growth arrest is far greater.

**Supracondylar Fractures**

Ossification centres are present in children. These are the sites of new bone formation. The presence of six ossification centres around the elbow can make interpreting a child's X-ray difficult (Figure 6). These ossification centres appear in a fairly constant order and can be remembered by the acronym CRITOE:

- **Capitellum** - age 1.
- **Radial head** - age 3.
- **Internal epicondyle** - age 5.
- **Trochlea** - age 7.
- **Olecranon** - age 9.
- **External epicondyle** - age 11.

Knowing the order of ossification is helpful. The most important reason is that the site of an ossification centre may be mistaken for a fracture. An example is shown in Figure 7.

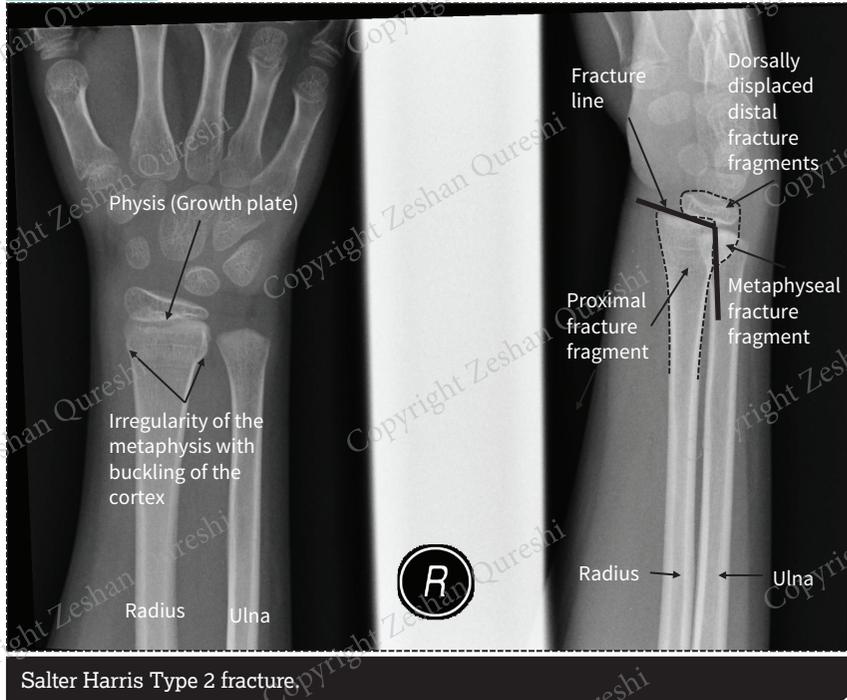
**Clinical Features**

Supracondylar fractures are common fractures in children. Classic features include:

- Arm extension injuries.
- They commonly occur following a fall onto an outstretched hand.
- Tenderness and swelling over the elbow.

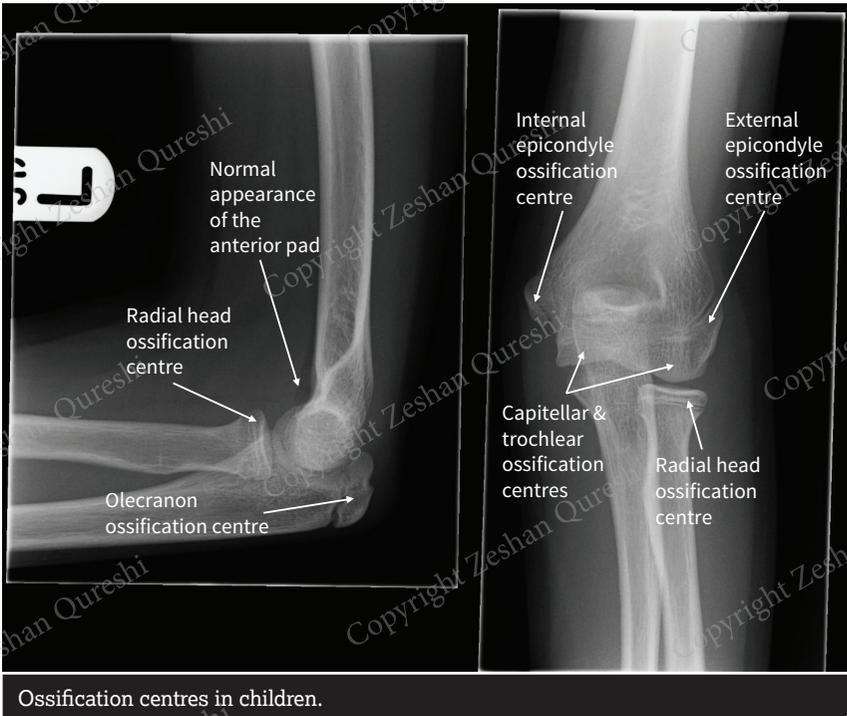
There is an associated forearm injury in 10%, and compartment syndrome may

FIGURE 5



Salter Harris Type 2 fracture.

FIGURE 6



Ossification centres in children.

ensue. There is also a risk of injury to the brachial artery and the radial, ulnar and median nerves.

**Management**

Management may be conservative, using plaster immobilisation, or

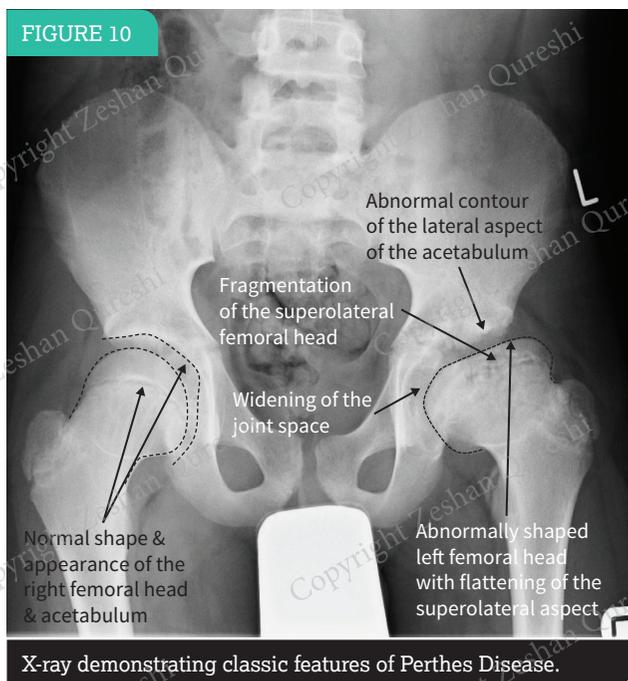
surgical, depending upon the degree of displacement and associated neurovascular injury.

**Clavicle Fracture**

This is the most common paediatric bone to be fractured (Figure 8).



FIGURE 10



X-ray demonstrating classic features of Perthes Disease.

### Differential Diagnosis

- ▶ **Transient synovitis.** This is a self-limiting condition in an otherwise well child.
- ▶ **Juvenile idiopathic arthritis.** This commonly affects more than one joint with extra-articular features.
- ▶ **Septic arthritis/Proximal femoral osteomyelitis.** Elevated inflammatory markers.
- ▶ **Proximal femoral fracture.** This follows significant trauma.
- ▶ **Acute or chronic slipped upper femoral epiphysis.** This is more likely in older, overweight children.

### Management

The aims of treatment are to relieve the symptoms, improve mobility, reduce mechanical stress and to contain the femoral head in the acetabulum until it reforms (containment). Treatment can be conservative or surgical and is governed by:

- ▶ The radiological severity of the disease.
- ▶ The degree of reduced hip motion.
- ▶ The age of the patient.

### Conservative Management

Mild to moderate disease (less than 50% involvement of the femoral head with preservation of joint space) is generally managed conservatively.

- ▶ The patient is initially kept non-weight bearing.
- ▶ Non-steroidal anti-inflammatory medications are used until pain subsides.
- ▶ Regular program of physiotherapy (once pain under control).
- ▶ If an adductor muscle contracture is present, botulinum toxin injections together with physiotherapy may help to improve containment by increasing the range of movement in abduction.

### Surgical Management

Surgical treatment may be required if there is severe disease, the patient is older than six, or when conservative management fails. If immobilisation in plaster cast is indicated, the majority of orthopaedic surgeons would recommend surgical management since immobilisation is heavily restrictive. Options include a varus osteotomy of the femur or innominate osteotomy of the pelvis. Both approaches aim to increase the containment of the femoral head in the acetabulum. A total hip replacement at skeletal maturity can be performed for patients with severe functional impairment.

### Complications

- ▶ Joint stiffness.
- ▶ Limb length discrepancy.
- ▶ Coxa magna. Broadening of the head and neck of the femur.
- ▶ Osteoarthritis. This is secondary to abnormal weight bearing.
- ▶ Premature fusion of the growth plates leading to overgrowth of the trochanter.
- ▶ Hinged abduction.

### Prognosis

In patients less than 6-years-old, the outcome is good, irrespective of treatment. Children more than 6-years-old with severe Perthes disease have significantly better outcomes with surgical treatment than those with conservative treatment.

However, half of all patients require joint arthroplasty after a median of 50 years.

Indices for poor prognosis include:

- ▶ Girls (earlier skeletal maturity).
- ▶ Onset beyond the age of 6-years-old (lower remodelling potential).
- ▶ >50% involvement of the femoral head.
- ▶ Lateral displacement of the femoral head impinging on the acetabular margin.
- ▶ Subluxation.

## BONE AND JOINT INFECTIONS

### Septic Arthritis

Septic arthritis is a serious infection of the joint space and an important cause of joint swelling in children. The most commonly affected joint in the paediatric population is the hip joint, although almost any joint can be affected. Monoarticular septic arthritis is classical, although infants may present with multiple joint involvement. Prompt diagnosis and treatment of septic arthritis is paramount to prevent rapid destruction of the articular cartilage and joint space.

### Aetiology

Infection can spread to the joint in three ways:

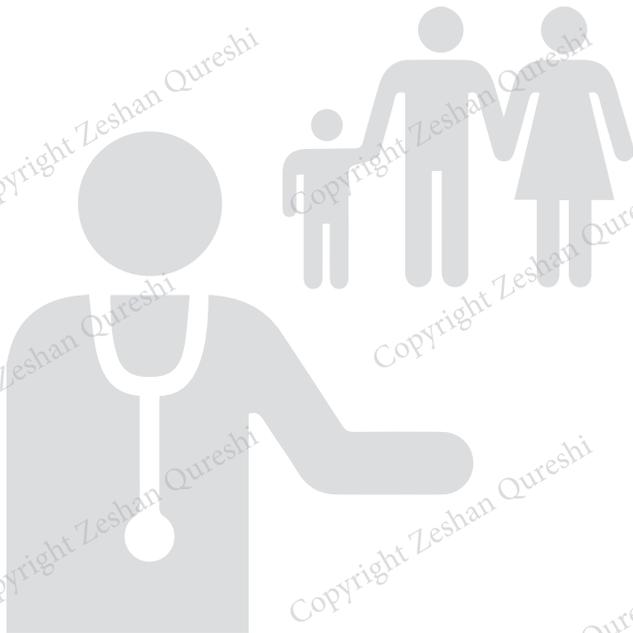
- 1 Direct invasion from an adjacent focus of infection, e.g. acute osteomyelitis.



# 1.20

## PUBLIC HEALTH

CHRISTOPHER HARRIS



### INTRODUCTION

Calculations for worldwide childhood morbidity and mortality are integral to healthcare planning and political direction. A 2013 article in the *Lancet* reported that 6.3 million children were estimated to die before the age of five, with 51.8% dying of infectious causes and 44% dying in the neonatal period. When compared to mortality rates in 2000, advances in treating pneumonia, diarrhoea and measles were collectively responsible for a reduction of approximately 1.8 million deaths. Projected estimates from this data predict 4.4 million children dying under five years in 2030. Part of the global health agenda is to identify and reduce preventable deaths.

Today, mortality is falling, but morbidity is increasing. For example, of the estimated 1.15 million babies with neonatal encephalopathy in 2010, 287,000 died, but a further 412,000 were estimated to survive with neurological impairment. With the provision of modern medicine and suitable intensive care, more babies are surviving, but the resultant medical complications continue throughout their childhood and into adulthood. These complications can reduce quality of life, and even if well managed, represent a major financial cost. More generally, children who would have died from acute infections or due to complications of a medical condition are surviving longer with chronic health concerns.

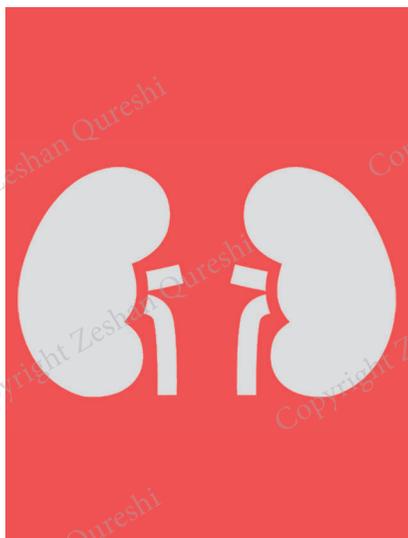
A summary of key causes of mortality, and morbidity are shown in *Table 1-2*.

The cost of providing healthcare worldwide is increasing. Ill health and disease have financial, social and potentially health implications that go beyond the single patient who seeks medical attention. Through public health, it is possible to reduce these burdens of healthcare and better manage the finite resources of individual regions, countries and the world as a whole. Forward planning, health promotion and service provision must evolve to meet the growing and ever changing demands of our diverse populations.

As we entered the new millennium, it was recognised that a common framework was needed to provide measurable improvements in health and wellbeing across the developing world. In September 2000, world leaders signed up to the United Nations Millennium Development Goals (MDGs). These were goals designed to focus efforts on a national and international level until 2015. Both progress and solutions

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# 1.21

## RENAL MEDICINE

ZESHAN QURESHI, RACHAEL MITCHELL  
AND STEPHEN MARKS

### CLINICAL MANIFESTATIONS OF RENAL DISEASE

#### Acute Kidney Injury

Acute Kidney Injury (AKI) can be defined as a sudden decrease in renal function; elevated urea is followed by an elevated creatinine and a resulting decreasing glomerular filtration rate (GFR). There are often associated difficulties in fluid and electrolyte regulation as well as with blood pressure control.

There are numerous definitions for AKI, but one which is widely used is the paediatric RIFLE (pRIFLE) which uses change in creatinine clearance and/or urine output for the definition (*Table 1*). Note that creatinine clearance is an approximation for the GFR, but is an overestimate (as creatinine is both filtered by the glomerulus, and secreted by the proximal tubule).

#### Aetiology

The causes of acute kidney injury can be pre-renal, renal or post-renal and are summarised in *Table 2*.

#### Clinical Features

Acute kidney injury can present nonspecifically, particularly in neonates with symptoms such as unexplained crying, restlessness, lethargy, vomiting, or poor feeding. Other features may include reduced urine output and pallor. Most commonly acute kidney injury is related to sepsis or hypovolaemia, in which signs of the cause would be apparent.

TABLE 1: Acute Kidney Injury definitions (pRIFLE)

Acute Kidney Injury	Creatinine clearance	Urine Output
Risk.	• Decrease by <25%.	• < 0.5ml/kg/hour for 8 hours.
Injury.	• Decrease by <50%.	• < 0.5ml/kg/hour for 16 hours.
Failure.	• Decrease by <75% (or <35mL/min/1.73m <sup>2</sup> ).	• < 0.3ml/kg/hour for 24 hours, or anuric > 12 hours.
Loss of function.	Prolonged loss of function (> 4 weeks).	
End stage.	Prolonged loss of function (>3 months).	

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- ▶ **Hypoalbuminaemia** (<25g/L).
- ▶ **Oedema.** May be a later sign or may not be present.

It is also usually associated with hyperlipidaemia.

### Aetiology

Eighty percent of cases in children are due to minimal change disease (Figure 1), which has normal light microscopy appearance, but a typical electron microscopy appearance with podocyte foot process effacement. Children usually present between 2 and 6-years-old. Rarely, children under 12-months-old can present with infantile nephrotic syndrome, or over 10-years-old with congenital nephrotic syndrome. These have a poorer prognosis.

Other less common causes are as follows:

- ▶ **Focal and segmental glomerulosclerosis.** Scarring in scattered regions of the kidney is seen

on histopathology. "Focal" means that only some of the glomeruli become scarred. "Segmental" means damage affects only part of an individual glomerulus.

- ▶ **Membranous nephropathy.** There is thickening of blood vessels in the glomeruli resulting in proteinuria and nephrotic syndrome.
- ▶ **Membranoproliferative glomerulonephritis.** This is a group of disorders. Some of them involve deposition of antibodies in the glomeruli, causing thickening and damage, which usually has a nephritic picture.

Risk factors include:

- ▶ **Asian ethnicity.**
- ▶ **Male gender.**
- ▶ **Previous infections.** Upper respiratory tract infections (URTIs) or hepatitis.
- ▶ **Medications.** e.g. NSAIDs (nonsteroidal anti-inflammatory drugs).
- ▶ **Systemic lupus erythematosus.**

- ▶ **Diabetes mellitus.**
- ▶ **Human immunodeficiency virus.**
- ▶ **Family history of nephrotic syndrome.**
- ▶ **Previous nephrotic syndrome.**

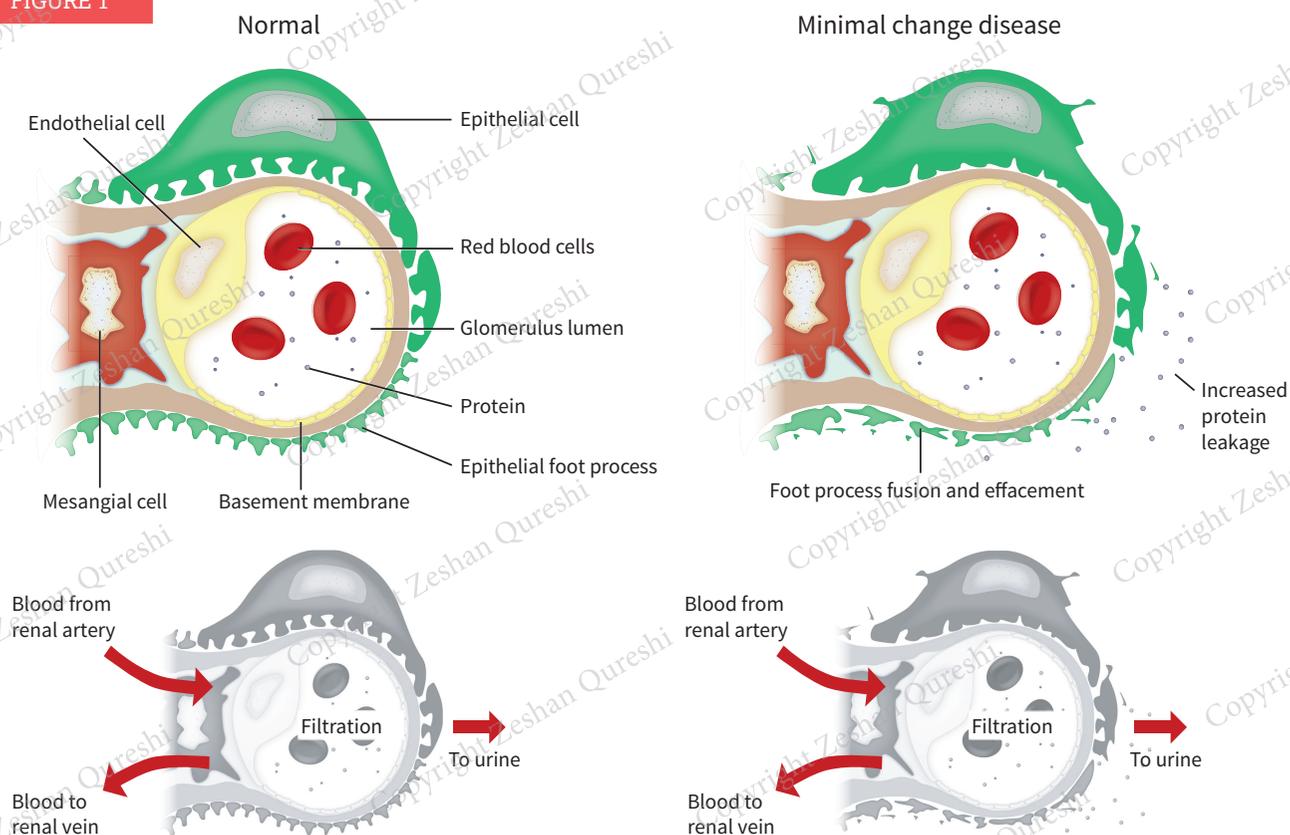
### Clinical Features

Symptoms vary depending on the underlying aetiology and the severity of the condition. It may simply present with mild oedema. Periorbital oedema is often the earliest sign (Figure 2) and may be initially misdiagnosed as an allergic reaction.

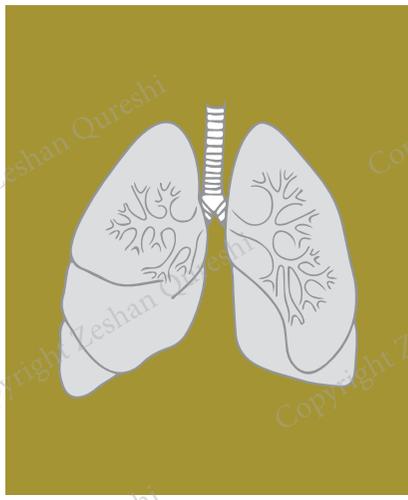
Other possible symptoms include:

- ▶ **Discomfort relating to swelling or skin breakdown.**
- ▶ **Weight gain.**
- ▶ **Abdominal distension.**
- ▶ **Tiredness.**
- ▶ **Foamy urine.**
- ▶ **Increased infections.** This is particularly from encapsulated organisms like pneumococcal or *Haemophilus* infection.
- ▶ **Poor growth and development.**

FIGURE 1



Minimal change disease.



# 1.22

## RESPIRATORY MEDICINE

CHRISTOPHER GRIME

### RESPIRATORY DISORDERS

#### Asthma

Asthma is a clinical diagnosis characterised by reversible airflow limitation.

#### Aetiology

Three main features give rise to the asthmatic phenotype:

- ▶ **Airway hyperresponsiveness.** This is an exaggerated bronchial smooth muscle contraction to a wide range of stimuli. The triggers vary according to the individual, but may include cold air, house dust mites or smoke.
- ▶ **Bronchial inflammation.** There is widespread inflammation in the bronchioles, with infiltration by eosinophils, T lymphocytes and mast cells. This is associated with oedema, smooth muscle hypertrophy, mucus plugging and epithelial damage. Some of these changes may be chronic but they are more pronounced during an asthma exacerbation.
- ▶ **Airflow limitation.** This is usually reversible, either spontaneously or with treatment, but there may be some underlying chronic changes.

Asthma is generally characterised by classical helper T cell type 2 (Th2) pathology with increased cytokines such as interleukin 4, 5 and 13 (IL-4, IL-5 and IL-13), which are thought to drive symptoms. The pathology of asthma is characterised by goblet cell hyperplasia and infiltration of inflammatory cells such as CD4<sup>+</sup> T cells, eosinophils and mast cells.

#### Clinical Features

As with most conditions, a clear clinical history is often all that is needed, particularly with young children. Symptoms are induced by weather changes, ill health and exercise. Nocturnal symptoms and improvement on bronchodilation are even more suggestive of the disease.

- ▶ **Wheeze.** Classically, in asthma, the wheeze is an expiratory airflow sound resulting from narrowed and inflamed airways. It can be heard at the bedside or require auscultation. However, the volume of the sound can be misleading. A

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# 1.23

## SKIN CONDITIONS

MAANASA POLUBOTHU

### OVERVIEW OF THE SKIN

**T**he skin is the largest organ in the body. Its key functions include:

- ▶ **Barrier function.** It prevents entry of water and electrolytes into the body through the surface.
- ▶ **Immunosurveillance.** Langerhans cells act as antigen-presenting cells. Macrophages and lymphocytes defend against microorganisms that breach its protective barrier.
- ▶ **UV protection.** Melanocytes contain the pigment melanin, which filters UV radiation from sunlight.
- ▶ **Thermoregulation.** This is orchestrated by sweat glands and blood vessels of the dermis, in addition to the insulating properties of the subcutaneous layer.
- ▶ **Sensory function.** Nerve endings detect touch, pain and temperature.
- ▶ **Vitamin D synthesis.** Vitamin D is converted into its active form in the basal and spinous layers of the epidermis under the influence of ultraviolet light from sunlight.
- ▶ **Respiration.** The skin accounts for up to 2% of respiration by absorbing oxygen and eliminating carbon dioxide.

There are three layers to the skin: the epidermis, dermis and the subcutaneous layer (*Figure 1*).

#### Epidermis

This is the thin, uppermost layer acting as a waterproof physical barrier. It is the first line of defence against the environment. The epidermis is immensely complex. It has four main layers: stratum basale, stratum spinosum, stratum granulosum and stratum corneum. Another layer, the stratum lucidum, is found on the palms and soles. The main cells of the epidermis are the keratinocytes. Other important cells are melanocytes (dendritic cells in the basal layer, containing melanin pigment) and Langerhans cells (dendritic antigen-presenting cells forming part of the immune surveillance system of the skin). The dermo-epidermal junction (DEJ) is the region that lies between the epidermis and the dermis holding it together. It is immunologically and genetically important; antibodies against various components of the DEJ are found in genetic and acquired skin conditions such as blistering diseases.

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FIGURE 4



Seborrhoeic dermatitis (D@nderm).

and intertriginous areas. Lesions seen in the neck folds and axillae tend to be confluent, erythematous and greasy, but non-scaling. Seborrhoeic dermatitis may also manifest as “cradle cap”, which favours the vertex and frontal areas of the scalp, in a “cap like” distribution (Figure 4). Pruritus, if experienced, tends to be mild. Infants generally thrive, with normal feeding and sleep.

#### Management

Parents should be offered reassurance and educated about simple skin care measures; the application of emollients, frequent shampooing and removal of scales with gentle brushing.

#### Prognosis

Most cases spontaneously resolve within a few weeks to months. Cases persisting for greater than 12 months may need further evaluation.

### Psoriasis

#### Aetiology

Psoriasis is a common skin disorder affecting two percent of the population. The pathophysiology is not fully understood; an altered immune response and polygenic inheritance are thought to play vital roles. Although psoriasis can occur at any age, it is relatively

uncommon in children, with only 10% of cases presenting before 10-years-old.

#### Clinical Features

Psoriasis is characterised by intensely erythematous, well-demarcated, symmetrical plaques with adherent silvery scales. Common sites are the elbows, knees, lumbar region, ears, scalp and hairline (Figure 5). Psoriasis in children can present on

the face. Guttate psoriasis is a form of psoriasis triggered by a throat or skin streptococcal infection. It presents suddenly, with widespread small plaques measuring up to 1 cm on the trunk and extremities.

#### Management

Management varies depending on disease severity and can include:

- ▶ Emollients.
- ▶ Vitamin D analogues.
- ▶ Tar preparations.
- ▶ Corticosteroids.
- ▶ Dithranol.

Second-line treatments include:

- ▶ Phototherapy.
- ▶ Systemic agents, such as retinoids, methotrexate or acitretin (in a minority of children).
- ▶ Biologic treatment with etanercept is now licensed for use in severe, refractory childhood psoriasis.

#### Complications

Associations include psoriatic arthritis in up to 48% of patients. This can affect the large or small joints but has a tendency to involve the distal small joints and may present with dactylitis. Additionally, nail changes such as pitting or onycholysis

FIGURE 5



Psoriasis plaques (D@nderm).

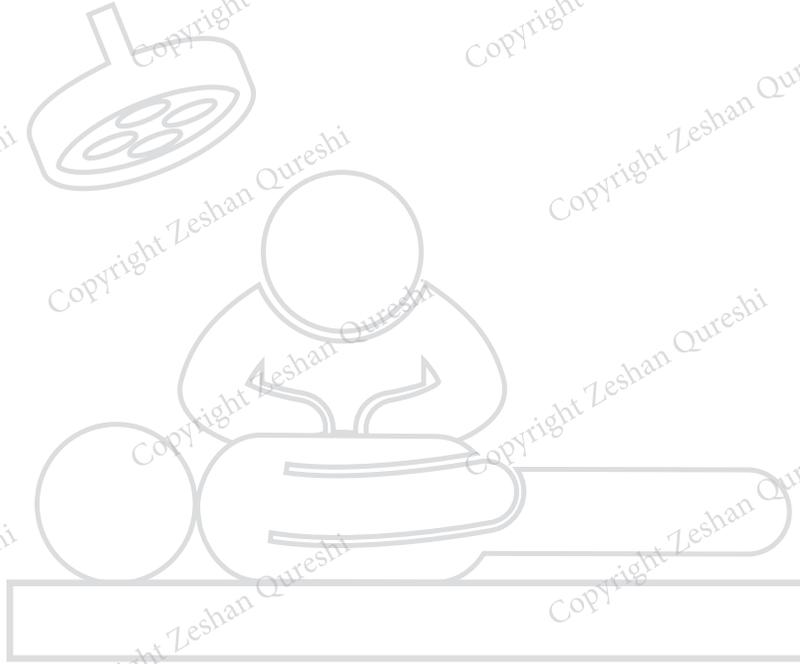




# 1.24

## SURGERY

MAY BISHARAT



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FIGURE 3



examined between bouts of pain, a sausage-shaped mass can frequently be felt in the right upper quadrant. Occasionally children have an extremely distended abdomen or can have frank peritonitis (rigid abdomen) and sepsis.

### Investigations

Blood tests may point towards inflammation, electrolyte derangement, dehydration and significant bleeding. However, the gold standard investigation is an ultrasound scan of the abdomen. The classic sign is the "target sign" (or doughnut sign) on transverse section, which represents the two concentric lumina of bowel (*Figure 3*). An abdominal X-ray may reveal features of small bowel obstruction.

### Management

#### Non-Operative Management

Appropriate fluid resuscitation should occur prior to any intervention as children with intussusception will have suffered huge third space losses of fluid. IV antibiotics (broad spectrum, like co-amoxiclav) should be administered promptly.

Following adequate resuscitation, a pneumatic reduction enema is performed:

- ▶ A catheter is inserted into the rectum of the child in the radiology department and air is insufflated under fluoroscopic screening.
- ▶ The pressure generated by the air, in 75-80% of cases, is sufficient to achieve a complete reduction (*Figure 4*).

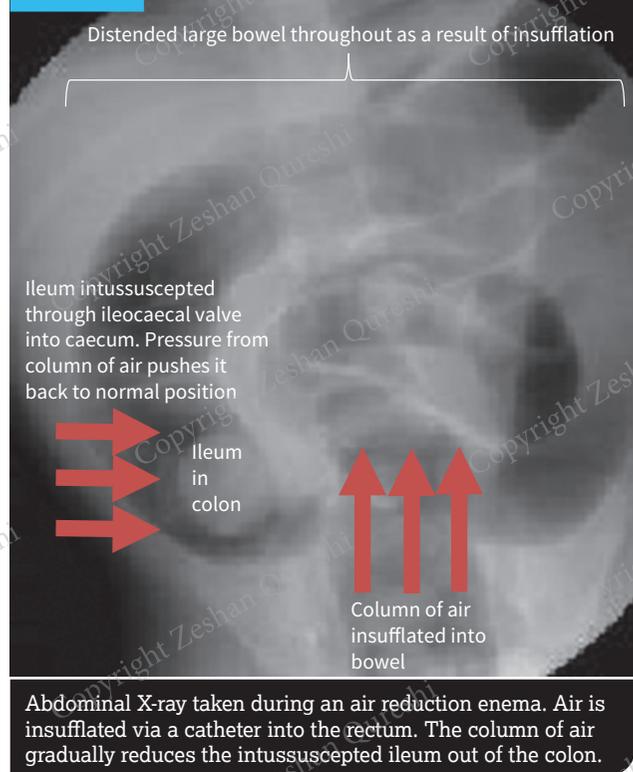
Patients who fail air enema, have a highly distended abdomen, or who are peritonitic will require an urgent laparotomy.

#### Operative Management

A right transverse incision is made and the intussusception is identified. In 50-60% of cases, a simple manual reduction is possible. In the remainder of cases, the bowel is either too oedematous to allow a successful reduction or has become non-viable. In such cases, a resection of the affected bowel with end-to-end anastomosis is performed.

Following a successful air reduction enema, fluids can be reintroduced after 12-24 hours and patients are discharged

FIGURE 4



48 hours later. Patients who undergo an open manual reduction have a similar post-operative course, whereas children who undergo bowel resection have a slightly longer recovery period.

### Complications

If left untreated, intussusception can lead to:

- ▶ Ischaemia.
- ▶ Necrosis.
- ▶ Haemorrhage.
- ▶ Perforation.
- ▶ Infection/peritonitis.

Complications can also be iatrogenic, with one percent of air reduction enemas leading to perforation.

### Prognosis

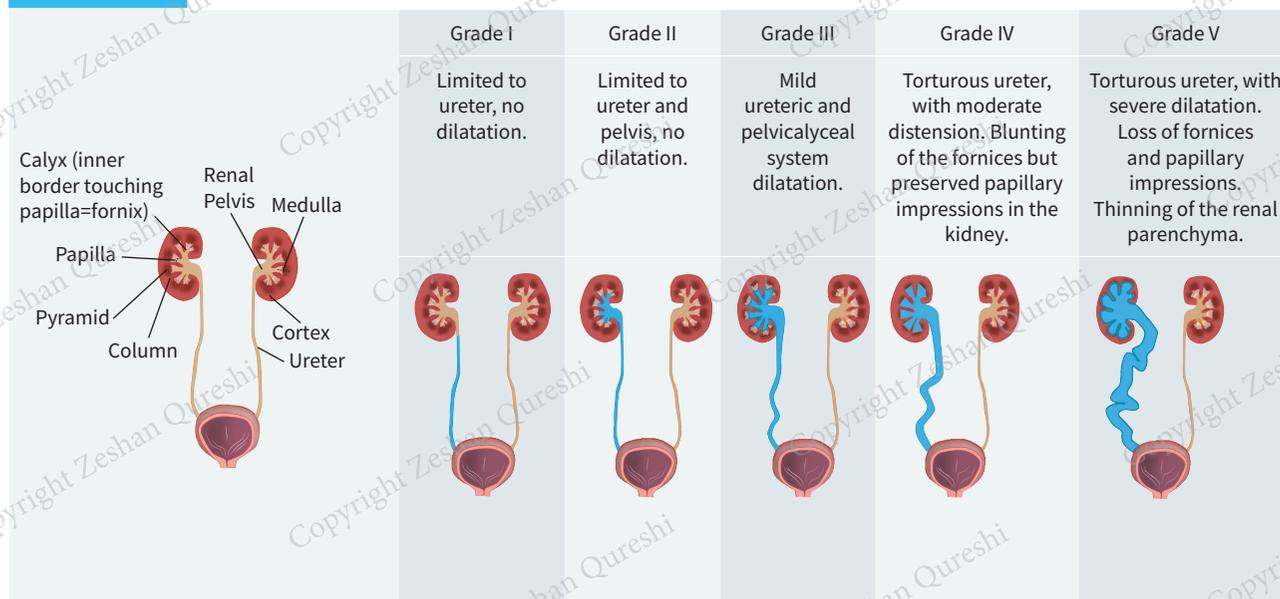
Mortality is approximately one percent and is mainly related to inadequate fluid resuscitation or diagnostic delay. Recurrence occurs in 10% of cases. Approximately 30% occur in the first 24 hours post reduction and 70% present within six months.

In children with more than one recurrence, investigations for a pathological lead point need to be performed.

### Inguinal Hernia

An inguinal hernia is the protrusion of peritoneal cavity contents through the inguinal canal. The incidence of inguinal hernias is approximately one to two percent, but is much higher in pre-term infants (up to 20%). The male to female ratio is four to one. The condition is more common on the right, although 10% of cases are bilateral.

FIGURE 8



Grading of VUR by the International Reflux Study Committee (1981).

TABLE 6: Aetiology of vesico-ureteric reflux

Primary VUR	Abnormally situated ureteric orifice. Larger than normal orifice. Short submucosal tunnel.
Secondary VUR	Obstructive: posterior urethral valves, meatal stenosis. Neuropathic bladder: spina bifida, spinal injury. Dysfunctional bladder.

Progressive renal scarring leads to loss of function. This, in turn, leads to renal failure and hypertension. Up to 15% of all cases of VUR develop end-stage renal failure.

The grades of VUR reflux are summarised in **Figure 8**. Grade I and II resolve relatively quickly, but the higher the grade, the greater the chance of renal scarring.

**Clinical Features**

VUR can be picked up:

- ▶ Most commonly, during the investigations arising after a UTI (p304).
- ▶ As part of the work up for antenatal hydronephrosis.
- ▶ Incidental findings (e.g. the finding of a scarred atrophic kidney on ultrasound for other reasons).
- ▶ Patients present with hypertension and/or end stage renal disease (rarely).

**Investigations**

MCUG is the gold standard for diagnosing reflux. Contrast is injected into the bladder, and images are taken during micturition (bladder contraction). Any reflux can subsequently

be seen. DMSA (dimercaptosuccinic acid) scanning is a nuclear medicine imaging modality that is very useful for the detection of any renal scarring. Hydronephrosis (distension and dilatation of the renal pelvis and calyces) can be seen on ultrasound (**Figure 9**).

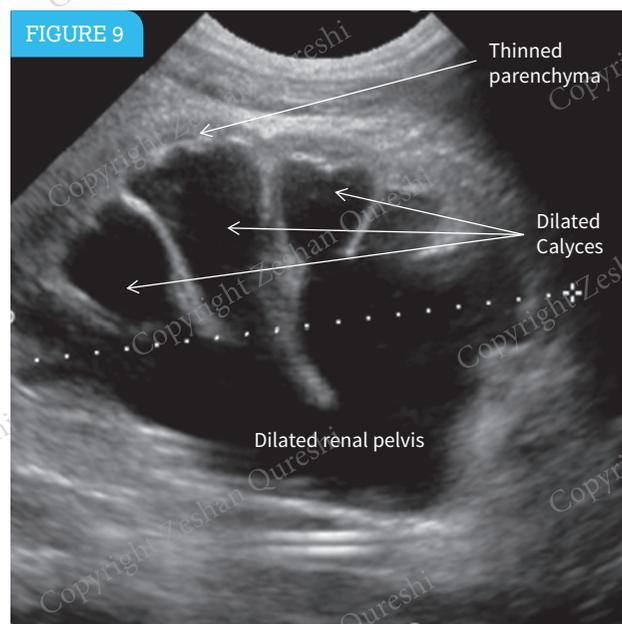
**Management**

**Medical**

The aim is to treat ongoing UTIs and prevent any further episodes. Medical treatment options for VUR include:

- ▶ Prophylactic antibiotics.
- ▶ Laxatives (to treat any associated constipation).

FIGURE 9



Ultrasound scan of kidney, showing hydronephrosis. There is dilatation of the pelvicalyceal system, and thinning of the renal parenchyma.



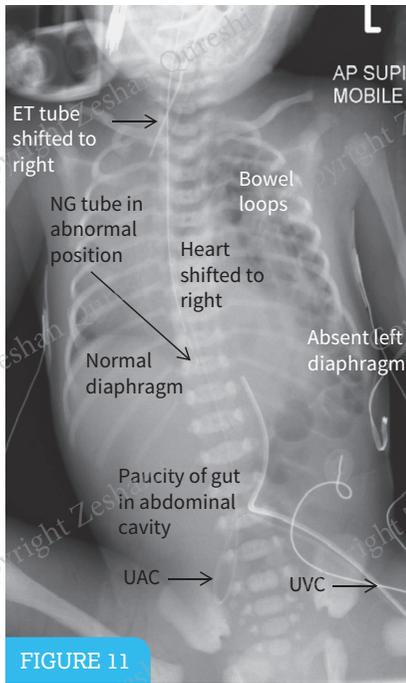


FIGURE 11

Chest and abdominal X-ray demonstrating a congenital diaphragmatic hernia. Loops of bowel are scant in the abdomen and extend into the left chest cavity. The mediastinum is shifted to the right, as demonstrated by the ET tube and the heart being on the right hand side. The NG tube may be insufficiently advanced as it is only just extending below the right diaphragm. The left diaphragm cannot be seen because of the degree of herniation.

involves insertion of a NG tube (to prevent distension of the stomach) and intubation/ventilation.

Surgical repair of the diaphragmatic defect is only performed once the infant is in a stable condition. The procedure begins with a subcostal incision and the defect is then repaired using remnants of muscle (primary repair) or prosthetic material (patch repair). Both laparoscopic and thoracoscopic CDH repairs are now performed.

In recent years, foetal surgery has undergone some trials but it is not yet widely adopted. Inserting a balloon into the foetal trachea to promote lung expansion is currently reserved for cases with poor outcome indicators.

**Complications**

Patients who are well enough to undergo CDH repair show some long-term sequelae:

- ▶ Respiratory insufficiency secondary to the pulmonary hypoplasia.
- ▶ Neurological and developmental issues secondary to hypoxia in the newborn period.
- ▶ Gastro-oesophageal reflux disease. This might be so severe that medical anti-reflux treatment is insufficient and a fundoplication is required.
- ▶ Chest wall and spine deformity. Some require surgical correction.
- ▶ Recurrence of the diaphragmatic hernia. This is more common following use of a synthetic patch.

**Prognosis**

Despite advances in antenatal and neonatal care, the mortality of the condition remains approximately 60%. The major determinant of survival is pulmonary hypoplasia, while presence of associated cardiac defects increases mortality to over 90%. Antenatal diagnosis gives a worse prognosis, as it is associated with a significantly greater lung hypoplasia.

**Tracheo-Oesophageal Fistula/Oesophageal Atresia (TOF/OA)**

In most instances, oesophageal atresia and tracheo-oesophageal fistulae occur together. The incidence of this condition is around one in 4000 live births.

In oesophageal atresia (OA), a portion of the mid-oesophagus is missing. The gap between the two remaining ends can be short (more common) or long (less frequent).

A tracheo-oesophageal fistula (TOF), by contrast, is a communication between the oesophagus and the trachea. This can occur as an isolated defect (an H-type fistula), but is very rare. The most common variant is a blind ending upper pouch of oesophagus, with the lower oesophagus connecting to the trachea; this is called a “distal” pouch fistula. This variant accounts for more than 85% of TOF/OA cases (Figure 12).

**Management**

If the condition was diagnosed on antenatal scans, delivery should be planned at a centre that provides neonatal intensive care and paediatric surgery. Neonatal resuscitation

FIGURE 12

Type 1	Type 2	Type 3	Type 4	Type 5
Isolated oesophageal atresia.	Proximal fistula with distal atresia.	Proximal atresia with distal fistula.	Double fistula with intervening atresia.	Isolated fistula (H-type).
8%	2%	85%	<1%	4%

Classification of TOF/OA.





# 2.01

## CLINICAL CASES: STANDARD

ALL AUTHORS

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CASE 1

**A 14-year-old girl presenting with self-harm**

**J**ennifer, a 14-year-old girl, is brought in by her mum after having self-harmed by cutting her arm with a razor. On assessment, the cuts are shallow and horizontal, with no active bleeding. The girl is quiet and withdrawn, but appears otherwise well. This is her first episode of self-harm. When asked whether she wanted to kill herself, she shrugs. Observations are within normal limits. After being reassured that her cuts do not require further treatment, both Jennifer and her mother are keen to go home.

Questions

- Q1** Which of the following would be the best way to manage Jennifer?
- A Jennifer should be sectioned because she is suicidal
  - B She should be discharged home in her mother's care, because she is a child
  - C She should be assessed before discharge by the child and adolescent mental health team, either in the ED or as an inpatient
  - D She should be discharged on her own because she has been assessed as Gillick competent
- Q2** When you speak to Jennifer alone, she tells you she has a 16-year-old boyfriend that her mother doesn't know about. On questioning, she admits that they are having sex but that "he doesn't like condoms". Which of the following is/are correct? Select all that apply.
- A You have a duty to inform the police because she is having underage sex

- B You should encourage her to talk to her mother about her boyfriend
- C You have a duty to tell her mother that she is having sex because she is under 16, but you don't need to tell Jennifer before you do this
- D If Jennifer has been assessed as Gillick competent, you do not need to do anything about this
- E If she requests it, Jennifer can be prescribed the pill without telling her mother

- Q3** You want to give Jennifer further advice regarding sexual health. Which of the following is/are correct? Select all that apply.
- A Jennifer should go on the pill so that she doesn't need to use condoms
  - B She should not be given any contraception, as this is encouraging her to have sex underage
  - C A coil is the best choice for her because she is young and hasn't had children yet
  - D She can go on her own for a sexual health screen
  - E Telling her about all the different methods of contraception will be confusing for her

For answers see page 390

CASE 2

**A 3-year-old boy presenting with stridor**

A 3-year-old boy is brought in by ambulance to ED, as his parents were concerned about his breathing. Yesterday, he had a hoarse voice, runny nose and temperature, but awoke this morning very distressed, with a barking cough and



**CHART 1A: BLANK PRESCRIPTION CHART FOR CLINICAL CASE 14**

<b>Name</b>			<b>Weight</b>			<b>Known allergies</b>		
<b>Date of birth</b>			<b>Gestation</b>					
<b>Hospital number</b>			<b>Consultant</b>					
<b>Admission date</b>			<b>Ward</b>					
<b>Signature:</b>			<b>Date:</b>					
<b>REGULAR MEDICATIONS</b>								
			<b>Date Time</b>					
<b>Drug</b>			<b>02</b>					
<b>Dose</b>	<b>Freq</b>	<b>Route</b>	<b>06</b>					
			<b>10</b>					
<b>Start</b>	<b>Stop/review</b>		<b>14</b>					
<b>Signature</b>			<b>18</b>					
<b>Indication</b>			<b>22</b>					
<b>Drug</b>			<b>02</b>					
<b>Dose</b>	<b>Freq</b>	<b>Route</b>	<b>06</b>					
			<b>10</b>					
<b>Start</b>	<b>Stop/review</b>		<b>14</b>					
<b>Signature</b>			<b>18</b>					
<b>Indication</b>			<b>22</b>					
<b>FLUID PRESCRIPTIONS</b>								
<b>Date</b>	<b>Fluid</b>	<b>Additive</b>	<b>Volume</b>	<b>Route</b>	<b>Rate</b>	<b>Signature</b>	<b>Given</b>	<b>Batch</b>

He is febrile, with focal crepitations on his chest. You suspect a lower respiratory tract infection. He has not been eating or drinking. He is to be admitted to Puffin ward under Dr Ross.

**Question**

Please write up a drug chart (Chart 2a) for him to include:

- ▶ Ten puffs of salbutamol inhaler every four hours
- ▶ An appropriate choice of oral antibiotics
- ▶ 2/3 maintenance fluids
- ▶ 30mg oral prednisolone once per day

For answers see page 407

**CASE 16**

**A 12-year-old girl presenting with abdominal pain**

A 12-year-old girl presents with a 36-hour history of vague central abdominal pain that has shifted to the right iliac fossa.

She is otherwise well. Her periods started four months ago but have been irregular: her last one was one week ago. She has vomited twice since the onset of pain, and complains of dysuria. Whilst waiting to be seen, the child had a bout of watery diarrhoea.

On examination, her HR is 120 bpm (tachycardic) and her temperature is 37.8°C, but all other observations are within normal limits. She is coryzal. She is tender over both sides of her abdomen.

**Questions**

Q1 Which of the following conditions is on your list of differential diagnoses? Select all that apply.

- A Urinary tract infection
- B Acute appendicitis
- C Ovarian pathology
- D Nonspecific abdominal pain
- E Gastroenteritis





## ANSWERS

## ANSWER TO CASE 1

## A 14-year-old girl presenting with self-harm

**Q1** Which of the following would be the best way to manage Jennifer?

The correct answer is C. She should be assessed before discharge by the child and adolescent mental health team, either in the ED or as an inpatient.

*When dealing with a child that has presented with self-harm, it is important to get an accurate assessment of their risk of further self-harm or completed suicide. A formal assessment is required by the child and adolescent mental health team before discharge. Their role is to make a discharge plan to keep the child or young person safe.*

- A** Jennifer should be sectioned because she is suicidal – Incorrect. Although this is a possibility, it would be very rare to need to section a patient following self-harm. This patient appears to be cooperative and not an immediate risk to her own safety, so there are other options that are more appropriate.
- B** She should be discharged home in her mother's care because she is a child – Incorrect. Children and young people should generally be admitted following a significant episode of self-harm. This could be admission to a paediatric or adolescent ward, if available. In some circumstances, review by the mental health team may occur in the ED, and if a plan is put in place for a safe discharge, this can be followed, but it is not the norm.
- C** She should be assessed before discharge by the child and adolescent mental health team, either in the ED or as an inpatient – Correct. Usually patients are only discharged from the ward after being formally assessed by child and adolescent mental health services. This may happen in the ED, depending on when the young person attends. If an assessment is not available on the same day, she should be admitted until she is assessed. It is also important to consider child protection concerns, and it may be appropriate to inform social care.
- D** She should be discharged on her own because she has been assessed as Gillick competent – Incorrect. As above, she should be admitted. Irrespective of her Gillick competence, she cannot refuse treatment that is deemed in her best interests.

## Key Point

Ensure all children presenting to hospital with self-harm have a formal specialist assessment of their risk of suicide before discharge.

**Q2** When you speak to Jennifer alone, she tells you she has a 16-year-old boyfriend that her mother doesn't know about. On questioning, she admits that they are having sex but that "he doesn't like condoms." Which of the following is/are correct?

The correct answers are B, You should encourage her to talk to her mother about her boyfriend and E. If she requests it, Jennifer can be prescribed the pill without telling her mother.

*When reviewing a child or young person, it is important to be aware of any possibility of abuse or exploitation. However, it doesn't necessarily follow that a relationship such as this is harmful. Sharing such information has to be balanced against the autonomy and confidentiality of the patient. Although Jennifer should be encouraged to openly discuss important issues with her parents, if there are no specific concerns about this relationship beyond age, it would be an unnecessary breach of confidentiality for the parents to be informed without explicit consent.*

- A** You have a duty to inform the police because she is having underage sex – Incorrect. Although she is under 16, if she is having consensual sex with someone close to her age, the police do not need to be informed. However, if abuse or exploitation is suspected, social services need to be informed. If she was under 13, this would always be reported, regardless of the nature of the relationship.
- B** You should encourage her to talk to her mother about her boyfriend – Correct. It is good practice to encourage young people to confide in their parents, even if they are reluctant. However, this cannot be forced. Be aware that, within some cultures, this may be particularly difficult for the young person or, in extreme cases, it may put the young person in harm's way.
- C** You have a duty to tell her mother that she is having sex because she is under 16, but you don't need to tell Jennifer before you do this. – Incorrect. This should only be done if there are concerns about the child's safety. If this was the case, it is good practice to inform a young person before breaking confidentiality.
- D** If Jennifer has been assessed as Gillick competent, you do not need to do anything about this – Incorrect. Although she may be Gillick competent relative to this decision, it is an important opportunity to give advice and any treatment required.

- E If she requests it, Jennifer can be prescribed the pill without telling her mother. – Correct. If she is Gillick competent, she can have this medical treatment confidentially.

### Key Point

Although safeguarding concerns are important, do not forget about the right to confidentiality of a child or young person. It should only be broken if doing so may prevent a clear, significant harm.

**Q3** You want to give Jennifer further advice regarding sexual health. Which of the following is/are correct?

The correct answer is D. She can go on her own for a sexual health screen.

Seeing a young person in the Emergency Department provides a good opportunity for further health promotion. Jennifer should be fully informed about her options with regard to contraception, including how to access contraceptive services. Barrier methods are important if there is any risk of sexually transmitted infections.

- A Jennifer should go on the pill so that she doesn't need to use condoms – Incorrect. She may choose to start taking the pill, but condoms are recommended in addition, to reduce the risk of sexually transmitted diseases.
- B She should not be given any contraception as this is encouraging her to have sex underage – Incorrect. There is no evidence to back up this statement, and it is important to promote “safe sex”.
- C A coil is the best choice for her because she is young and hasn't had children yet – Incorrect. Although coils are a very effective form of long-acting reversible contraception, it may be slightly more difficult to insert a coil in younger patients. As such, contraceptive implant use is preferable.
- D **She can go on her own for a sexual health screen – Correct. This makes sexual health services much more accessible to adolescents.**
- E Telling her about all the different methods of contraception will be confusing for her – Incorrect. Giving Jennifer all of the information about available forms of contraception will enable her to make the most appropriate choice for her. This gives her autonomy over her own care and increases the chances that she will use contraception effectively.

### Key Point

Just as with an adult patient, with adolescents, the duty of a doctor involves ensuring patients have the full information required to make informed decisions.

### IMPORTANT LEARNING POINTS

- When dealing with self-harm, it is important to assess the risk of further self-harm or suicide. This should be followed by referring or sign-posting other services that are available to support the young person. These can be general (for example, community mental health teams) or specific to an individual problem (for example, drug addiction).
- Sexually active adolescents have a right to access contraceptive services. They should be fully informed of their options, including how to access care.
- Encourage sexually active adolescents to have a sexually transmitted disease screening, if they haven't done so already.
- It is important to be very aware of the possibility of abuse or exploitation in any relationship. If this is suspected, then it is appropriate to break confidentiality in the best interest of the patient.

## ANSWER TO CASE 2

### A 3-year-old boy presenting with stridor

**Q1** What is the most likely diagnosis?

The correct answer is E. Croup.

The five options given are the five commonest causes of stridor, which should be considered in the differential diagnosis of any case with stridor.

- A Epiglottitis – Incorrect. Stridor is present, but children with epiglottitis appear unwell, with high fevers and drooling.
- B Bacterial tracheitis – Incorrect. Unlike children with croup, these children are toxic, with rapidly progressive airway obstruction. Onset is not as rapid as with epiglottitis. Bacterial tracheitis is rare but very serious. It can be difficult to differentiate clinically from epiglottitis, but the treatment is the same.
- C Anaphylaxis – Incorrect. Anaphylaxis is associated with an urticarial rash, wheeze, shock, and often vomiting and diarrhoea. This patient has no clear precipitant for anaphylaxis and none of the other features.
- D Inhaled foreign body – Incorrect. A barking cough and infective symptoms make this unlikely. However, sudden-onset respiratory distress can be a sign of foreign body inhalation, particularly in a young child. It is important to ask about the possibility of foreign body inhalation specifically in the history.
- E **Croup – Correct. The progressive nature of the symptoms and worsening overnight are classical of croup. The patient is interacting well, with only a low-grade fever, and is not tachycardic. All these features suggest that, despite presence**





# 2.02

## CLINICAL CASES: INTERMEDIATE ALL AUTHORS

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**449 Case 19**

A 2-year-old presenting with a blue episode

**449 Case 20**

A 5-week-old girl presenting with respiratory distress and poor weight gain

**450 Case 21**

A 4-year-old boy presenting with nephrotic syndrome

**450 Case 22**

An 11-year-old boy presenting with haematuria

**451 Case 23**

A 5-year-old girl presenting with a purpuric rash

**CASE 1**

**A 5-year-old girl presenting with an acute neck swelling**

**A** previously fit and well 5-year-old girl is reviewed in ED with a progressively enlarging, left-sided neck swelling. She reports having a sore throat and has a fever of 38°C, but observations are otherwise normal.

**Questions**

**Q1** What clinical feature(s) below would be of concern and prompt further investigation?

- A Night sweats
- B Coryzal symptoms
- C Hepatosplenomegaly
- D Stridor
- E Weight loss

**Q2** On examining the child, what finding(s) would suggest acute infection? Select all that apply.

- A Overlying erythema
- B Multiple, bilateral cervical lymph nodes.
- C Solitary, painless neck mass
- D Fluctuant mass
- E Fever

**Q3** If a non-infective pathology is suspected, which three of the following should be organised as first-line investigations?

- A MRI scan of the neck
- B Ultrasound scan
- C Blood film
- D FBC
- E Excision biopsy

**Q4** With regard to neck masses in children, mark the following statements true or false.

- A The most common histological feature on biopsy is reactive hyperplasia

- B Most palpable lymph nodes over 0.5 cm will require an excision biopsy
- C Branchial cysts most commonly present in the neonatal period
- D Supraclavicular swellings are common in children
- E Neck swellings are rarely associated with infectious mononucleosis

For answers see page 452

**CASE 2**

**A 10-year-old boy with newly diagnosed diabetes**

Jamie is a sporty and active 10-year-old. His mother noticed that recently his clothes have seemed too big for him, and he was always complaining of feeling tired. Jamie was very embarrassed to admit that he had wet the bed on two occasions this week. Observations are within normal limits.

On examination, Jamie is very thin, with signs of recent weight loss. Jamie's urine dipstick shows 4+ glucose, 2+ ketones, 1+ leucocytes and is negative for nitrites.

A new diagnosis of diabetes is suspected.

**Questions**

**Q1** Which of the following best fits with a diagnosis of Type 1 diabetes in this child?

- A A high blood glucose, only if it is seen following a formal oral glucose tolerance test
- B A random blood glucose >11.1 mmol/L
- C The presence of polyuria and polydipsia
- D 4+ of glucose on urine dipstick
- E The presence of ketonuria

**Q2** Regarding the aetiology of diabetes, mark the following statements true or false.

- A All diabetes in children is type 1
- B Type 1 diabetes mellitus is an autoimmune condition caused by damage to pancreatic alpha cells





# 2.03

## CLINICAL CASES: DIFFICULT

ALL AUTHORS

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### CASE 1

#### A 2-year-old girl presenting with knee pain

**H**ayley is a 2-year-old girl who has been brought to ED by her parents. She banged her right knee on a table two weeks ago. It was sore at the time, but she has subsequently mobilised on it. She has been unable to weight-bear on her right leg for the last 48 hours and she has a swollen right knee. Her HR is 155 (tachycardic) and her temperature is 38.5°C, with observations otherwise within normal limits. She has been off her food for the last day and “hasn’t been herself”, according to her parents. Initial bloods show a white cell count of  $13.1 \times 10^9/L$ . On examination, her right knee is markedly swollen and is held in a flexed position (around 60°). She is reluctant to flex or extend the knee due to pain.

### Questions

- Q1** What should be the working diagnosis?
- A Fracture
  - B Knee ligament injury
  - C Septic arthritis
  - D Transient synovitis
  - E Osgood-Schlatter disease
- Q2** What additional investigations might be helpful in establishing a diagnosis? Select all that apply.
- A X-ray right knee
  - B CRP
  - C ESR
  - D Ultrasound right knee
  - E X-ray right hip
- Q3** What is the next best stage in management of this patient?
- A Immediately commence broad-spectrum antibiotics
  - B Admit and observe the patient
  - C Immediately commence broad-spectrum antibiotics and then aspirate the knee





# 3.01

## HISTORY TAKING

ZESHAN QURESHI, CHRISTOPHER HARRIS  
AND MICHAEL MALLEY

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### INTRODUCTION

**W**hen approaching a potentially unwell child, it is important to remember that children are not just small adults. Bear in mind the following:

- ▶ The child's parents, relatives or carers may not have the whole story, especially if the child has been in someone else's care, so a collateral history may be necessary.
- ▶ The parents' "sixth sense" about their child's health should always be taken seriously. Equally, parents may underestimate the seriousness of their child's condition.
- ▶ Younger children may not be able to give any history, or the history given may be misleading.
- ▶ All children, but particularly younger infants, may have underlying congenital abnormalities that have remained undetected until presentation in extremis.
- ▶ Children can compensate physiologically for severe illness. As such, signs of deterioration may not be evident until late in the illness course.
- ▶ Children are dependent on adult carers for many things, particularly in early life. This can lead to opportunities for abuse, which can be easily missed if not specifically assessed.
- ▶ Decisions made during childhood with regards to health can impact on all areas of development.
- ▶ Pay special attention to putting the child at ease. The doctor should make a point of introducing themselves to the child individually, and perhaps let them lead introductions to the rest of the family. Toys are widely available in paediatric healthcare settings and they can be used effectively to relax younger children.

### PREPARATION

Usually, there will be time to prepare before seeing a child. During this time, review any pre-existing information. This may include: ambulance triage notes, nursing triage notes, referral letters, previous clinic letters, hospital discharge letters and current hospital notes. Often, allergies or social concerns are automatically highlighted on hospital computer notes. There may also be a handover from a colleague: don't be afraid to ask them questions to clear uncertainty. Blood tests and imaging may have been requested, or recent investigation results may be available. This will give an initial impression of the key issues and the parent/patient agenda before going into the consultation.



## CASE 3: A 13-YEAR-OLD BOY PRESENTING WITH A WHEEZY EPISODE

### Social History

Question	Justification	Answer	Evolving thought process
Who is currently at home? Does Fred have a social worker? Is dad around? Does anyone smoke at home?	Find out if there are any social concerns, or if wider support is needed. Smoking increases risk of infection in the child.	"My husband and I live together, and have been happily married for several years. We are both non-smokers, and there is no social worker."	No social concerns at present.

Immunisation history is not relevant in a six-week-old baby, as the first immunisations are at eight-weeks-old.

### Concluding

Question	Justification	Answer	Evolving thought process
Is there anything that I've missed, or anything that you are particularly concerned about? Do you have any questions for me at the moment?	May identify useful information that has been missed. Will also help framing communication with the mother.	"I am really keen to go home."	Needs a careful and sensitive discussion about a management plan.



### PRESENTING YOUR FINDINGS

Fred is term baby who presents at six weeks of age very well in himself, but with a one-off temperature at home of 39°C.

There is no difficulty in breathing, no seizures, no rashes, no vomiting and no diarrhoea. Fred is feeding well at the breast, with a normal number of wet nappies.

Jamie was born by an elective C-section for breech, required no resuscitation and went home with mum after a normal newborn baby check. Mum had one raised temperature during labour, and a background of Group B streptococcus, with two doses of antibiotics given during labour. Antenatal scans are normal. Fred is growing well, tracking the 50<sup>th</sup> centile. Grandma and granddad both currently have an upper respiratory tract infection (URTI), but there is no family history of note. This is mum's second child, and the other child is well. There are no social concerns.

I am concerned about possible infection, particularly considering the maternal background, although there is no clear focus for infection identified.

After doing a full examination, I will cannulate the baby, and check the FBC, U&Es, CRP and blood cultures. I will get a clean catch urine sample, and a chest X-ray if there are any signs of respiratory disease on examination. A nasopharyngeal aspirate will be helpful if the baby has signs of possible bronchiolitis. In view of the history of Group B streptococcus and fever, I would also perform a lumbar puncture. I will start broad spectrum antibiotics. Fred is currently feeding well, so I'd be happy for him to continue breastfeeding at present.

### CASE 3

#### A 13-year-old boy presenting with a wheezy episode

#### Doctor Briefing

You are asked to see a boy, Steven, 13-years-old, in the ED who has started wheezing at home. Please take a history from his mother, with a view to making a diagnosis.

#### Mother (actress) Briefing

Steven is your only child, and you are worried about him because he is wheezing and has difficulty breathing. He is breathing fast and having to pause in the middle of sentences to catch his breath. His shortness of breath has come on gradually. He does not have a fever but he has had a bit of a cold for the last few days. He is off his food, but drinking about two-thirds of normal. Steven occasionally has had wheezy episodes in physical education lessons, but he usually gets better quickly.

Steven is otherwise well, is not on any medication and has never attended hospital before. He was born at 35 weeks by a normal vaginal delivery. He is up-to-date with his immunisations, developing normally, growing well and performing well academically.

You are a single mother, but are coping well. Steven stays with Dad every other weekend. You don't smoke. Dad and the paternal granddad both have eczema.

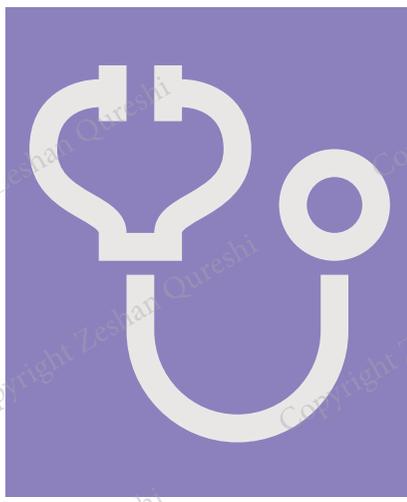
You are concerned because this is the worst you've ever seen Steven, and he can't talk to you properly. If specifically asked, you are also worried that his dad smokes and that this might contribute to the breathing problems.

#### Assessment

For all scenarios:

- Introduction.
- Ask a mixture of open and closed questions.
- Ensure understanding throughout.
- Summarise your findings to the parents, and give them an opportunity to raise any other issues.





# 3.02

## EXAMINATION

AMY MORAN AND ZESHAN QURESHI

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to examine lots of patients, including siblings of inpatients and young relatives. Practice will make it easier to combine clinical knowledge with examination findings to form a diagnosis – or at least it will draw some broad differentials.

The examination chapter is laid out in order of some of the key systems that feature in both acute, outpatient and exam settings. In nearly all circumstances all systems will need to at least briefly be assessed.

Certain rules apply to all examinations:

- ▶ Remember to wash hands before seeing a patient.
- ▶ Begin with an introduction to both the parent and the child.
- ▶ Explain what is going to be done before doing it.
- ▶ With younger children, take consent from parents. Tell the child what is going to be done. If the child is asked whether it is ok to examine their chest and they then say “no,” it is then difficult to proceed. Instead say, “now I am going to listen to your chest.”
- ▶ When examining for pain, look at the child’s face to gauge their reaction rather than the part of the body being examined.
- ▶ When finished, thank the child.

MEDICAL DEVICES

Children will often have medical devices in situ. A good knowledge of it may identify relevant past medical history, as well as facilitate ongoing management (e.g. knowing where blood samples might be taken from).

Central Venous Access

- ▶ **Portacath®.** This is an implanted port that sits under the skin connected to a central venous catheter (Figure 5). The port is accessed with a specific needle (Figure 6). Medication can be given via the port and blood can be taken from it. It is seen most commonly in patients undergoing long-term intravenous treatments, such as chemotherapy, regular blood transfusions (e.g. sickle cell disease, thalassaemia) or regular intravenous antibiotics (e.g. cystic fibrosis). The

FIGURE 1



main complications are infection, port dislodging (the port can sometimes flip too), thrombosis or blockage of the catheter.

- ▶ **Hickman line.** This is a brand of tunnelled central venous catheters used for long-term treatment such as chemotherapy (Figure 7). Medication may be given via the line and blood can be taken from the line. If not used regularly, the lines should be flushed. As with any central venous device, blockage, dislodgement, thrombosis and infection can occur. The line can also split; it may sometimes be possible to repair small splits depending on their location. Note that a **Broviac line** is similar to a Hickman line, but the lumen is smaller, meaning it can be used in smaller children and infants.
- ▶ **PICC (Peripherally Inserted Central Catheter) line.** These may be inserted under local or general

INTRODUCTION

**P**aediatric examinations can be fun and are a chance to make your patient smile and laugh.

They are a balance between systematic and opportunistic examination. Although some children can be unwilling or too upset to be examined, using play techniques can help win over any tantrum (Figure 1-4). For some children, this will be their first encounter with a doctor so keeping the interaction friendly helps to minimise fear. With older children, chat to them, find out their likes and dislikes and explain to them what you are going to do before you do it, at each stage. Wherever possible, remain at the same height as the child to reduce intimidation.

Practice is essential, particularly before a clinical exam. Use every opportunity



FIGURE 76



A



B

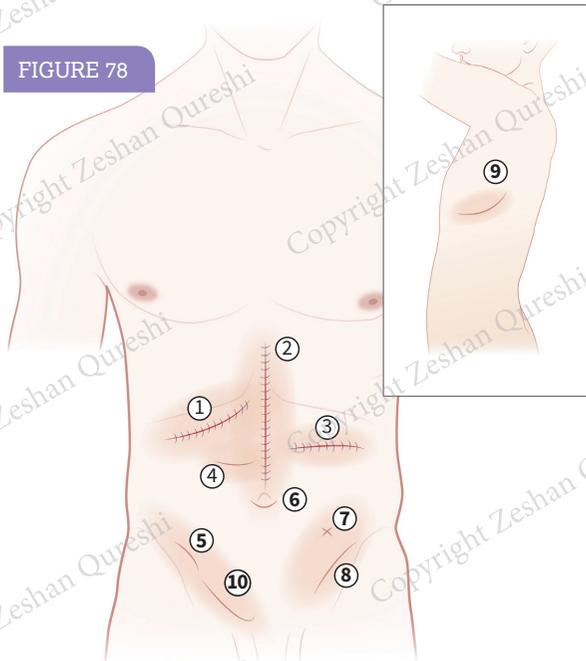
A. Exposing the sclera to look for jaundice. B. 10-year-old boy with jaundice.

FIGURE 77



Inspect the mouth, looking mainly at dentition and for ulceration.

FIGURE 78



Common abdominal scars in paediatrics.

▶ **Dentition.** Poor dentition is associated with malnutrition.

▶ **Glossitis.** Inflammation of the tongue. This is associated with iron, B12 or folate deficiency.

▶ **Angular stomatitis.** Inflammation of the corner of the mouth. May relate to iron, thiamine, or B12 deficiencies.

▶ **Freckling around the mouth.** This is associated with Peutz-Jeghers syndrome, which can present with GI bleeding from polyps.

▶ **Dysmorphic features.** Facial features can give a clue as to possible pathology in a syndrome. For example, Alagille syndrome is associated with a broad forehead, sunken eyes, small chin and chronic liver disease.

### Neck and Upper Trunk

▶ **Lymph nodes.** Palpate lymph nodes in the neck and supraclavicular fossa.

▶ **Gynaecomastia.** This may be from excess oestrogens in chronic liver disease. It is also a normal variant, particularly in pubertal boys.

▶ **Spider naevi.** This is a central red spot with extensions radiating out like a spider's web. It is due to dilated blood vessels, and occluding the central

spot will result in seeing the emptied peripheral veins filling from the centre. Classically seen in the superior vena cava distribution.

▶ **Central venous access devices.** There may be a temporary central line, e.g. post liver transplant, or a long term tunneled line, e.g. those on long term TPN. If they have since been removed, scars might be seen.

### Abdomen

Ensure that the child is lying flat with their arms by their sides. Ask them to point to the site of any pain. Inform the child that this will be examined last and

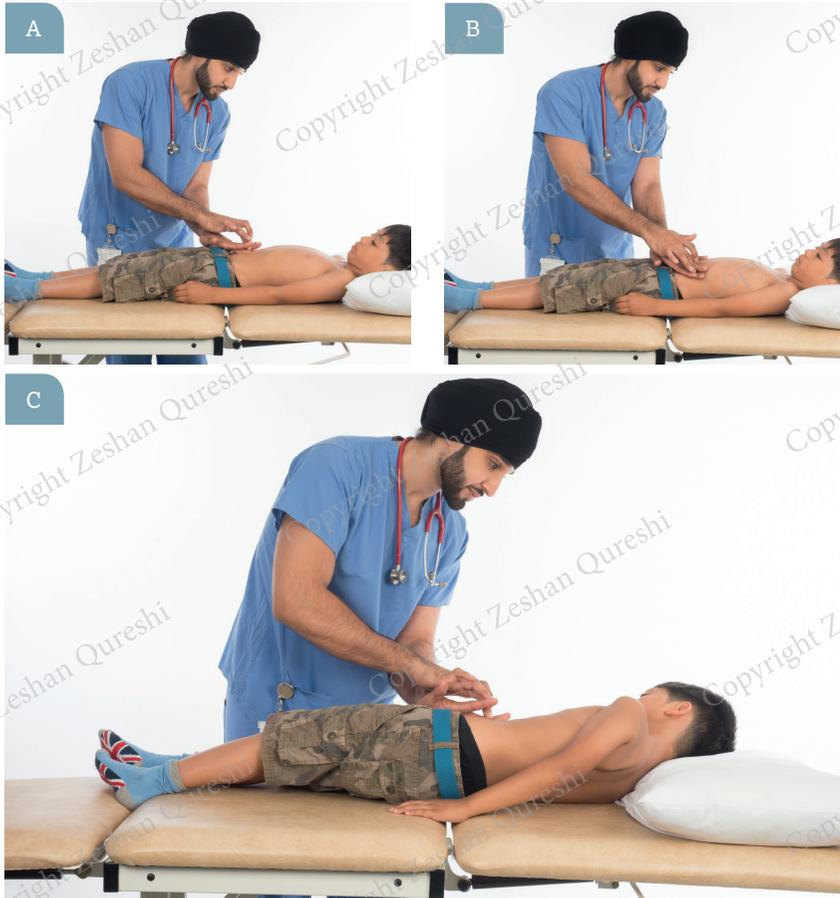
FIGURE 79



Transverse scar across the upper abdomen from a liver transplant.

- 1 **Kocher's incision**  
Biliary/liver surgery e.g. Kasai procedure for biliary atresia. Extended transversely for liver transplant.
- 2 **Midline laparotomy** Upper midline — upper GI surgery e.g. Nissen's fundoplication for GORD. Longer midline incisions are used for major abdominal surgery more generally.
- 3 **Transverse upper abdominal incision** Congenital diaphragmatic hernia repair. Splenic surgery.
- 4 **Pyloromyotomy scar** Pyloric stenosis surgery.
- 5 **Appendicectomy scar** Appendicitis surgery.
- 6 **Umbilical/sub-umbilical scar** Hernia repair, exomphalos repair. Gastroschisis repair has scar within/in place of the belly button.
- 7 **Point incision scar** Drains, ports, or biopsy sites.
- 8 **Inguinal scar** Hernia repair.
- 9 **Lateral thoracolumbar scar** Renal surgery e.g. nephrectomy.
- 10 **Hockey-Stick scar** Renal transplant.

FIGURE 84



Assess for shifting dullness as follows: A. Percuss from the centre of the abdomen outwards, until a dull percussion note is found (representing fluid). B. Pinpoint the location where the note becomes dull. C. At this point, the examiner should keep their hand in the percussed position, whilst rolling the patient towards them. Wait 30 seconds, and then re-percuss. If the note is resonant, this implies that the fluid has shifted under gravity. Confirm this by rolling the patient back into their original position and re-percussing. The percussion note should become dull again.

FIGURE 85



Assessing for a fluid thrill. Ask a parent to place a hand over the centre of the abdomen (to prevent conduction through the skin). Firmly flick the abdomen on one flank and feel for a 'thrill' on the other side.

▶ **Bladder.** The bladder is dull to percuss, and if enlarged, can be measured by percussion up from the suprapubic region.

**Auscultation**

Listen around 2 cm above the umbilicus:

- ▶ **Bowel sounds.** These may be normal, tinkling e.g. bowel obstruction, or absent e.g. peritonitis.
- ▶ **Renal bruit.** Present in renal artery stenosis.

**Completing the Examination**

Depending on the presenting symptoms, as well as performing a general paediatric examination, it is important to also inspect:

- ▶ **The anus.** Looking for fistula/skin tags (associated with Crohn's disease).
- ▶ **The external genitalia and hernia orifices.** Abdominal pain could be referred pain, e.g. from testicular torsion.
- ▶ **Rashes.** e.g. purpura (HSP), pyoderma gangrenosum and erythema nodosum (IBD).
- ▶ **Pitting oedema.** This may be from hypoalbuminaemia, e.g. chronic liver disease.

Bedside tests include:

- ▶ Review the observation chart.
- ▶ Plot the height and weight on an age and sex appropriate chart.
- ▶ Stool sample.
- ▶ Urine dipstick.
- ▶ Blood glucose.





## CASE SCENARIO

## SCENARIO 8

Claude is a 4-year-old boy, who attends the renal outpatient clinic for routine review. You are asked to perform an abdominal/renal examination on him, and present your findings to the consultant.



## EXAMINATION CHECKLIST

Domain	Findings	Evolving thought process
Global assessment of child on approach.	<ul style="list-style-type: none"> <li>Alert, active, and looks well.</li> </ul>	<ul style="list-style-type: none"> <li>Unlikely to have an acute pathology.</li> </ul>
Inspection.	<ul style="list-style-type: none"> <li>Previous central venous access scar.</li> <li>Gastrostomy in situ.</li> <li>Small scar to the right of his umbilicus.</li> <li>Midline laparotomy scar.</li> </ul>	<ul style="list-style-type: none"> <li>Has had major abdominal surgery in the past, which is likely to be the time when central venous access was acquired. Gastrostomy implies that there are current/past feeding difficulties.</li> <li>Scar to the right of the umbilicus may relate to a previous surgical drain.</li> </ul>
Palpation.	<ul style="list-style-type: none"> <li>Egg-shaped mass over midline.</li> <li>Laparotomy scar.</li> </ul>	<ul style="list-style-type: none"> <li>Likely transplanted kidney. Given this information, the scar to the right of the umbilicus may have been a site of peritoneal dialysis.</li> </ul>
Percussion.	<ul style="list-style-type: none"> <li>No organomegaly.</li> <li>No ascites.</li> </ul>	<ul style="list-style-type: none"> <li>No additional pathology identified.</li> </ul>
Auscultation.	<ul style="list-style-type: none"> <li>Normal.</li> </ul>	<ul style="list-style-type: none"> <li>No additional pathology identified.</li> </ul>
Completing the examination.	<ul style="list-style-type: none"> <li>Normal.</li> </ul>	<ul style="list-style-type: none"> <li>No additional pathology identified.</li> </ul>
Bedside tests.	<ul style="list-style-type: none"> <li>Awaiting.</li> </ul>	<ul style="list-style-type: none"> <li>Although looks well, possible faltering growth suggested by previous chronic disease (necessitating renal transplant), and gastrostomy being in situ.</li> <li>May have blood/protein in urine with any renal pathology.</li> </ul>

## MUSCULOSKELETAL EXAMINATION

A partial musculoskeletal or locomotor examination is undertaken in every paediatric assessment: gait is more often than not observed, as is an assessment of muscle bulk and gross movements. Consider the following more specific circumstances:

- In a child presenting with joint pain, stiffness, or loss of function (e.g. difficulty with handwriting). Is this an isolated joint pathology/injury or part of a wider problem?
- Screening for associated musculoskeletal involvement in a child with a predisposing condition. For example, psoriasis, IBD.
- Assessment of hypermobility.

Signs and symptoms of joint disease are shown in **Box 6**. Common patients that are likely to appear in exams are shown in **Table 18**.

## Box 6: Clinical features of joint disease

- Pain.
- Swelling.
- Stiffness.
- Erythema.
- Locking.
- Instability.
- Loss of ability/function.



## PRESENTING YOUR FINDINGS

Claude is a 4-year-old who was seen in the renal clinic for review.

He looks well. He has evidence of previous central venous access. He has a gastrostomy in situ, a small scar to the right of his umbilicus and a midline laparotomy scar. The abdomen is soft and non-tender throughout, with no clinically detectable organomegaly. There is a firm, non-tender, egg-sized mass underlying the laparotomy scar. There was no associated bruit. He has no other stigmata of disease.

These findings are consistent with a renal transplant. The scar to the right of his umbilicus is probably from previous peritoneal dialysis. It is not clear why he has a gastrostomy, but it could be for gastrointestinal or neurological pathology.

To complete my examination, I would like to review the observation chart, dipstick the urine and formally plot Claude on a growth chart.

## Summary Checklist

**Global assessment of the child on approach.**

**pGALS screening.** Gait, arms, legs, spine.

**Hypermobility screening.** Beighton score.

**Inspection.** Hands, skin, eyes, spine, individual joints.

**Palpation.** Each individual joint.

**Movement.** Passive then active movement of joint in question.

**Function.** For important activities e.g. doing up buttons.

**Completing the examination.** Auscultate heart (if hyperflexibility), palpate abdomen (if malignancy/inflammatory arthritis suspected).

**Bedside tests.** Review observations; plot height and weight.

FIGURE 122



Reinforcement.

In under 1:

- ▶ Primitive reflexes as described in Table 30.

**Sensation**

**Light Touch, Pain, and Temperature (Spinothalamic Tract)**

- ▶ Light touch, pain and temperature are all tested as described in the arms section. Dermatomes for the legs are shown in Figure 123.

**Proprioception and Vibration Testing (Dorsal Column)**

- ▶ **Proprioception.** Hold the big toe either side of the interphalangeal joint, and perform this test as shown with the arms. If proprioception is not intact at the interphalangeal joint, test it at the carpometatarsal joint, then the ankle, and then the knee.

- ▶ **Vibration sensing.** Vibrate a tuning fork and place it on the big toe, and perform this test as shown for the arm. If they cannot identify this, repeat on the malleolus, and then the tibial shaft, the tibial tuberosity, and finally the anterior iliac crest.

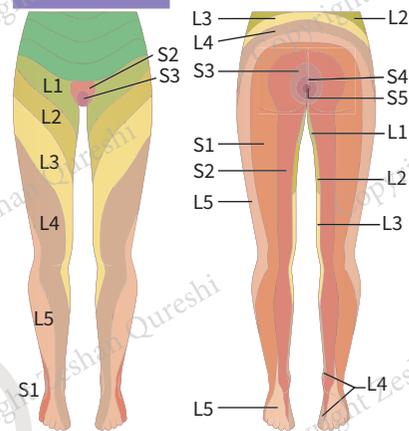
Sensation tests are summarised in Figure 124.

Peripheral nerves are less commonly damaged in the lower limbs than the upper limbs. However things to look out for are shown in Table 31.

**Co-ordination**

- ▶ **Heel-shin test.** Figure 125.

FIGURE 123



Dermatomes of the leg.

TABLE 31: Peripheral nerves of the lower limb

Nerve	Motor loss	Sensory loss
Obturator nerve (L2-4).	• Hip adduction.	• Medial thigh.
Femoral nerve (L2-4).	• Hip flexion. • Knee extension.	• Anteromedial thigh. • Medial lower leg, to medial malleolus.
Common peroneal nerve (L4-S3).	• Foot dorsiflexion loss gives characteristic foot drop. • Foot eversion loss gives inverted posture.	• Anterolateral lower leg. • Dorsum of foot/toes.
Sciatic nerve (L4-S3).	• Paralysis of all muscles below knee- gives foot drop from weight of foot (via tibial and common fibular branches).	• Posterolateral/anterolateral lower leg. Plantar surface of foot (tibial branch). • Lateral lower leg, and dorsal foot (common fibular branch).



FIGURE 124

Sensation testing, whilst the patient's eyes are shut. A. Light touch with cotton wool. B. Pain with a neurotip. C. Vibration sense with a tuning fork, vibrating. D. Vibration sense with a tuning fork, stopping the vibration. E. Proprioception in the "down" position. F. Proprioception in the "up" position.





# 3.03

## COMMUNICATION

ANNA CHADWICK AND MICHAEL MALLEY

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**611** Case 6: A 6-month-old boy who has not been vaccinated due to parental choice

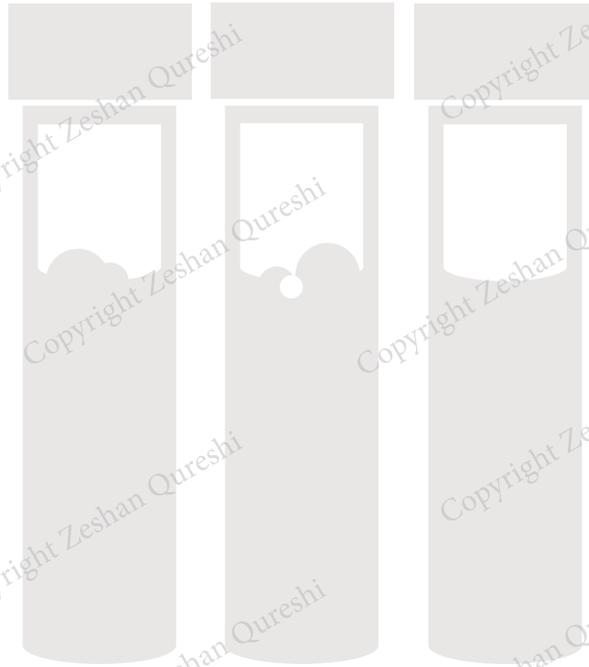
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# 3.04

## PRACTICAL SKILLS

MICHAEL MALLEY, ZESHAN QURESHI,  
ANITA DEMETRIOU AND MARIE MONAGHAN



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## INTRODUCTION

The key to success is preparation and practice. The best way to accomplish this is to discuss the procedure with a senior colleague and observe the skill in order to understand the process, before performing it under supervision. In addition to knowing the steps, it is important to know the indication for a procedure and its impact on management. This chapter contains a brief “how to” guide for each of the key competencies expected of junior doctors and paediatric trainees.

Important themes common to all procedures include:

- ▶ **Explanation.** Explain in lay terms what is going to be done, and why, to the child and parents. Invite questions.
- ▶ **Consent.** Gain informed consent and ensure the patient and parents are aware of any serious or common side effects. Consent may be written or verbal depending on the procedure.
- ▶ **Pain relief.** This may involve the use of oral/intranasal/intravenous/topical anaesthesia, cold spray, or oral sucrose.
- ▶ **Distraction.** Nearly all procedures are better tolerated with the use of appropriate distraction beforehand. Involve the play specialist team where possible. If unavailable, utilise smartphone apps/videos/toys as appropriate.
- ▶ **Aseptic technique.** Observe aseptic or sterile conditions as appropriate.
- ▶ **Keep calm.** Ensure adequate preparation, with appropriate support to hand. Being calm increases the likelihood of success. Know what to do in the event of any unexpected events.
- ▶ **Post-procedure.** After each procedure, clear the equipment, follow-up results and act on results as necessary.

A flow chart for performing procedures is shown in *Figure 1*.

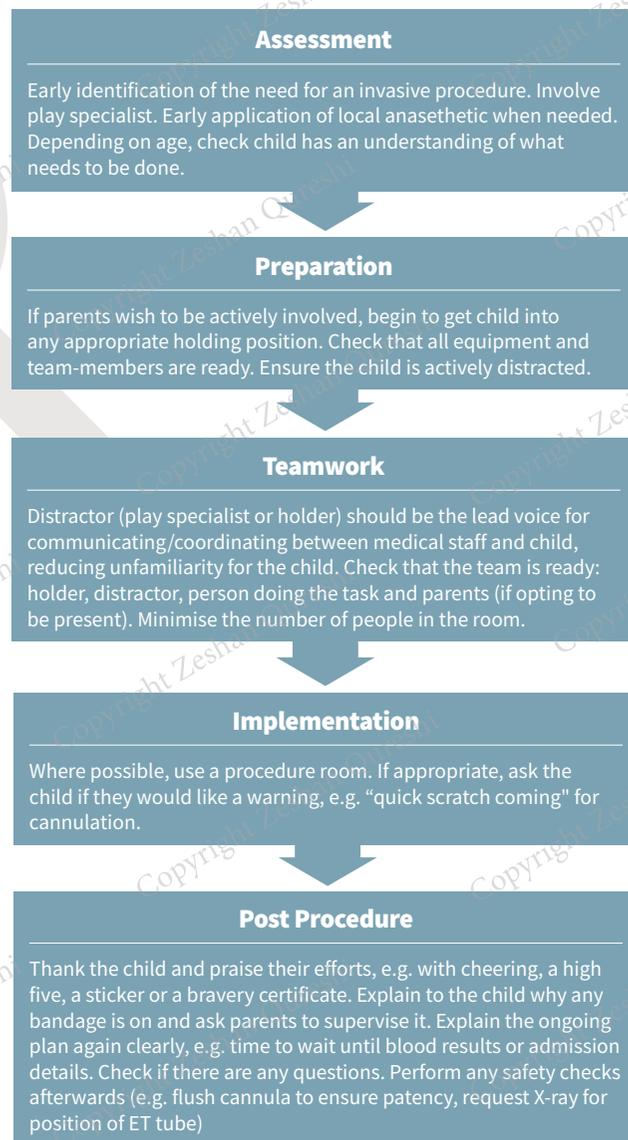
## VASCULAR ACCESS

### Capillary Blood Gas (“heel prick”)

Arterial blood gases are rarely performed in paediatrics unless from a secure arterial line in a high dependency setting. As a result, capillary gases fulfil an important role in the assessment of the ill child. Capillary blood gases are one of the most readily available objective barometers of a child’s health. They are quick and easy to obtain, with almost immediate results.

#### Indications

- ▶ Acutely unwell patient, e.g. respiratory compromise, severe sepsis.
- ▶ Checking glucose, bilirubin, haemoglobin or electrolytes.
- ▶ Post intubation or for assessing the efficacy of ventilation.



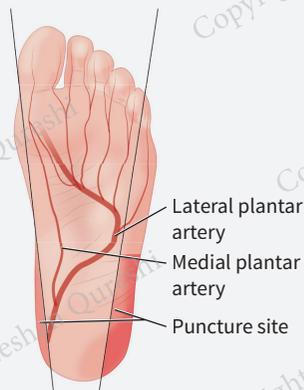
**FIGURE 1**

Flowchart for performing an invasive procedure.



**STEP-BY-STEP GUIDE**

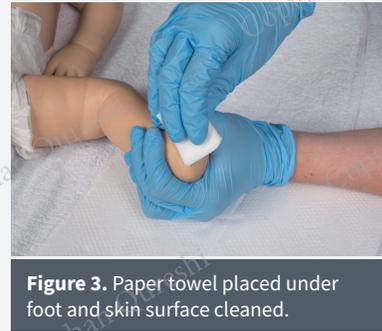
- 1 Introduce oneself to the patient/family and explain the purpose of the procedure.
- 2 Wash hands and put on gloves.
- 3 Identify the best place to perform the test:
  - › In a neonate, this will be a heel prick. This means selecting a spot around the periphery of the inferior aspect of the heel. Steer clear of the centre of the heel, where there are fewer capillaries and more nerve fibres (Figure 2).



**Figure 2.** Puncture sites for a heel prick. Avoid the centre of the heel.

- 4 Gently rub the site and/or gently push down at the intended site a few times to improve capillary blood flow. Check for any previous site of capillary blood sampling that will still bleed without requiring another piercing of the skin.
- 5 Place a paper towel under the intended puncture site to catch any spillages.
- 6 Clean the area and allow it to dry (Figure 3).
- 7 Apply a thin layer of Vaseline over the area to be punctured (note this is optional and not used in some neonatal units). This will ensure that the blood stays in a bleb around the needle prick site rather than smearing elsewhere, making it easier to collect.

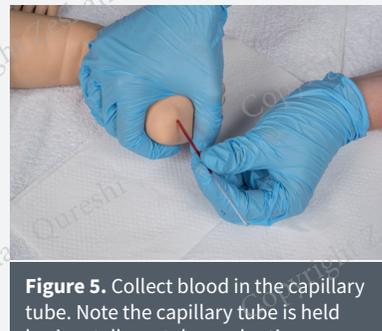
- 8 Take a mildly firm hold of the heel or digit and stretch the skin. Use the lancet to pierce the desired area (Figure 4).
- 9 Gently squeeze the area rhythmically, starting distally from the site of puncture and allowing reperfusion between squeezes. This should produce a steady bleb of blood from the site.
- 10 Hold the capillary tube horizontal or a little elevated. Holding it at a downhill angle may mean air bubbles are sucked in as the blood is collected quicker than it is produced. Holding it level or “uphill” from the patient will prevent this (Figure 5).
- 11 Gently move the end of the capillary tube to the bleb of blood and fill it to the end. Aim to only collect blood if it is free flowing so that clots are less likely to enter the capillary tube. If air bubbles enter, empty the capillary tube up until the point of the air bubble on gauze and refill the tube.
- 12 Once filled sufficiently, put the gas tube on a tray. Wipe the puncture site with some cotton wool or gauze to remove any remaining blood and any Vaseline (Figure 6).
- 13 Apply a plaster if needed (Figure 7). Note that this will not stick if Vaseline is still present.
- 14 If available, put a cap on both ends of the capillary tube as this will prevent spillage on transfer to the gas machine.
- 15 Thank the parents/patient and take the sample to the blood gas machine.
- 16 Remove the caps from the gas tube and put a clot catcher on the end to be inserted into the gas machine (if required).
- 17 Place lancet and capillary tube in a sharps bin.
- 18 Once the gas result has been obtained, immediately record it and make a plan based on the findings.



**Figure 3.** Paper towel placed under foot and skin surface cleaned.



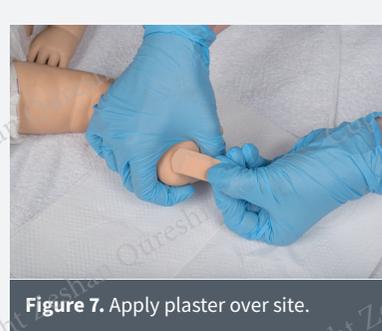
**Figure 4.** Tightened skin punctured with lancet.



**Figure 5.** Collect blood in the capillary tube. Note the capillary tube is held horizontally, not down-slanting.



**Figure 6.** Compress to stop bleeding with cotton wool.



**Figure 7.** Apply plaster over site.



# 3.05

## PRESCRIBING

MICHEAL MALLEY AND MARI MONAGON

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**648 Differences between Adults and Children**

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### THE IMPORTANCE OF GOOD PRESCRIBING

**G**ood prescribing is essential for safe, effective clinical practice. It remains, however, a common source of potentially harmful errors. This chapter will focus on the art of paediatric prescribing and discuss common pitfalls. Almost all errors may be avoided by being meticulous and thoughtful. Prescriptions will be checked by pharmacy and nursing staff before being administered, but the ultimate responsibility remains with the prescriber.

Remember:

- **Do not become complacent.** It is easy to feel that prescribing common medications is a mundane chore which does not require full attention. Try to remember that every prescription is a complex process. There is no shame in looking up drug doses and no honour in prescribing medications solely from memory.
- **Never prescribe an unfamiliar medication.** Do not prescribe a drug at the behest of a nurse or senior colleague, as ultimately the prescriber takes responsibility for it. Look up the drug, and know its indications and side effects before prescribing. If still unsure, seek advice from a senior doctor or pharmacist.
- **Always check the allergy status of the patient before prescribing.** This should always be filled out on the drug chart, and this simple step can prevent potentially fatal consequences.
- **Be aware of local and national protocols and know where you can access the dose, regimen, side effects and contraindications of each drug you prescribe.** In the UK, the British National Formulary for Children (BNFc) is the universal reference point, whilst individual hospitals will have their own specific guidelines.

### DIFFERENCES BETWEEN ADULTS AND CHILDREN

A number of subtle yet important differences exist between adult and paediatric prescribing. These are outlined below.

#### Drug Metabolism

Pharmacokinetics of medicines differs considerably in children and adults. For example:



# 4.01

## UNDERGRADUATE AND POSTGRADUATE ASSESSMENTS IN PAEDIATRICS

MICHAEL MALLEY AND MARIE MONAGHAN

### EXPECTATIONS OF A MEDICAL STUDENT

Although most medical students will not become paediatricians, doctors encounter children in most specialties, such as primary care, ENT and other surgical specialties, and emergency medicine, amongst others.

For undergraduates, senior clinicians want the student to approach the rotation in “good faith.” This means feeling that the student has tried their best, made the most of their time on the ward and shown a measurable improvement in performance. This is not demonstrated in a one-off assessment, but continually throughout the attachment. In terms of competence, it is important the student is confident that they:

- ▶ Are aware of the basic management and recognition of common paediatric problems.
- ▶ Appreciate the differences between the management of children compared to adults, including communication challenges.
- ▶ Understand the differences in adult and paediatric basic life support.
- ▶ Are aware of child protection issues and start to understand their management.
- ▶ Have completed the requisite range of assessments (which may require organisation on the part of the student).

Before a supervisor meeting at the end of the placement, it is important to think about the main learning points from the rotation, as well as what has gone well, and not so well. Demonstrate evidence of engagement with the children, and the paediatric team. Be aware that supervisors will usually take advice from other paediatric staff to make a holistic assessment of each student.

For postgraduates, educational supervisors have a long list of criteria to “tick off”. Become familiar with this list. Other factors to consider include evidence of quality improvement projects (e.g. audits), managerial experience (e.g. rota co-ordination), teaching experience, exam success and reflective practice. This should all be apparent in the trainee’s learning portfolio. More generally, remember assessments are increasingly recognised as learning opportunities. Prepare

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**Box 1: Presenting a history**

I met Mrs. Brown today, who brought her 7-year-old son Alex to ED with a 24 hour history of difficulty breathing and wheeze.

Alex is known to have asthma. He has had a cough for two days and has developed progressive difficulty in breathing and wheeze over the last 24 hours. He has a low grade temperature and a dry cough. He has been using two puffs of his salbutamol inhaler every four hours without improvement.

Alex has had many previous admissions to hospital, the most serious of which required IV salbutamol two years ago. His last admission was two months ago. His known asthma triggers are pets and upper respiratory tract infections, and he also has a history of eczema. His normal peak flow reading is around 300 mL/min. He misses about two days of school per month due to his asthma.

Alex was born at term without complication and there have been no concerns about his development.

Alex takes two puffs of a beclomethasone inhaler twice per day and uses his salbutamol inhaler when required. He takes no other medications. He has no known drug allergies and his immunisations are up-to-date.

Alex's mother has a history of asthma and his father has hay fever.

He lives in a flat with his parents and his 5-year-old brother. There are no smokers in the house and the family have never had any involvement with social services.

In summary, Alex is a 7-year-old boy who presents with a likely acute exacerbation of his asthma.

I would like to proceed to examine Alex, view his observations chart, perform a peak flow measurement and initiate bronchodilator therapy.

**Examination**

Examination stations cause anxiety amongst undergraduate and postgraduate candidates alike. What if the child isn't co-operative or cries? What if signs are missed? Candidates often think they will be heinously penalised for forgetting pallor or lymphadenopathy or missing grade one clubbing. They aren't! Remember that the overall conduct of the examination counts more than the sum of its individual minutiae. Be professional, sensitive, structured and observant. The examiner will be asking the question: *Is this person appropriate for the next level?* Students should act like a junior doctor; junior doctors should act like a middle-grade doctor.

**Before the Examination**

Practise. A lot. Become familiar with the important components of each examination, and practice under supervision with real patients. This will ensure technique is committed to muscle memory, making it easier to concentrate on picking up relevant clinical signs.

The first and last minute of each station are crucial. Therefore, practise the introduction and take time to put the patient at ease. Then practise presenting the examination findings in one minute. Practise presenting both real and imaginary patients. Practise to the mirror, to friends, and to colleagues.

Patients suitable for examinations nearly always have chronic conditions and are clinically stable. Contemplate what signs they may have and make a list of the common underlying diagnoses (*Table 2*). Ask senior students what they encountered in their exams. It may be helpful to create maps of common differential diagnoses to consolidate key findings. *Figure 1* is an example for abdominal examination.

**Starting the Station**

Do simple tasks well. Remember: the first minute is critical. Hands should

**TABLE 1: History taking mark scheme**

Domain	Mark				
<b>Conduct of the consultation.</b> To include: <ul style="list-style-type: none"> <li>• Introduction, clarifies role.</li> <li>• Rapport.</li> <li>• Empathy and respect.</li> </ul>	Clear Pass	Pass	Bare fail	Clear fail	Unacceptable
<b>History taking.</b> To include: <ul style="list-style-type: none"> <li>• Clear history of presenting complaint.</li> <li>• Appropriate style.</li> <li>• Fluent and comprehensive history.</li> <li>• Explores and responds to concerns/feelings.</li> <li>• Summarises and checks understanding.</li> </ul>	Clear Pass	Pass	Bare fail	Clear fail	Unacceptable
<b>Differential diagnosis and initial management.</b> To include: <ul style="list-style-type: none"> <li>• Appropriate range of differentials appropriate for age.</li> <li>• Accuracy of information/clinical knowledge.</li> </ul>	Clear Pass	Pass	Bare fail	Clear fail	Unacceptable
<b>FINAL GRADE.</b>	Clear Pass	Pass	Bare fail	Clear fail	Unacceptable



# 4.02

## A GUIDE TO BEING A JUNIOR DOCTOR IN PAEDIATRICS

MARIE MONAGHAN AND MICHAEL MALLEY

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Ward

### GENERAL PREPARATION

**P**aediatrics is a fulfilling and stimulating career, offering the potential to bring joy to children and their families. A unique specialty with its own diverse range of conditions, paediatrics requires a flexible approach, an appreciation for the breadth of medicine, and a dextrous hand for the wide range of procedures involved. As much as it demands, it rewards. Paediatricians laugh, play games and most certainly become familiar with the most popular Disney films and children's TV shows!

Although exciting for those already familiar with paediatrics, not surprisingly, those starting out can find this field incredibly daunting after an education focused on adult-centred medicine (*Table 1*).

Before starting a general paediatric or neonatal job, practitioners may benefit from attending a paediatric life support course. These are usually incorporated into paediatric induction training. Being confident in resuscitation will lead to increased confidence when facing the most demanding situations. This training can also lend a structured approach to the care of any child and provide advice on the most effective way to communicate and interact in resuscitation scenarios. Induction training will also offer a good opportunity to explore the specific working environment of the paediatric department.

*Table 2* shows the key areas in Paediatric units, and *Table 3* shows those areas that aid in coordinating patient care. Multidisciplinary teamwork is extremely important in paediatrics, meaning that effective communication is paramount for ensuring all parties are up to date with developments in a child's care. Being open with colleagues about previous levels of experience will help them tailor teaching, day-to-day task allocation and senior support.

The main chapters of the book give detailed information on key conditions encountered in paediatrics and key skills required for assessing and treating children. What follows is additional practical information to help with adaptation to life on paediatric wards.





# 4.03

## CAREERS IN PAEDIATRICS

MARIE MONAGHAN AND MICHAEL MALLEY

### EXPOSURE TO PAEDIATRICS

**P**aediatric clinical experience at medical school can be rather limited. As a result, those interested in paediatrics might have difficulty in attaining the desired amount of exposure to the specialty.

Approach an educational supervisor or a paediatric consultant about getting more experience in the paediatric ED or general paediatric ward. Often, these environments seem like chaotic places but be proactive and offer to clerk in patients for the paediatric team. This can contribute to history taking and can be a useful way to gain feedback.

Current junior doctors can arrange a “taster week”. This is a dedicated week of study leave arranged through an educational supervisor to give exposure to a speciality of interest. Talk to paediatricians, exploring what they love and hate about their career (*Table 1*). When on a placement, try to develop practical and communication skills. What may at first seem impossible, such as cannulating a 4-year-old, can become a highlight of the job. Note also that specialties like ED, primary care, ENT and many others come with exposure to paediatric patients.

Volunteer. Most cities have charities aimed at supporting young people. How about using a free afternoon to mentor a pupil? Volunteering will not only improve interaction skills with children, but will also generate meaningful experiences to draw on to support a paediatric speciality application. Incorporate paediatrics into a medical elective: this will provide an alternative perspective on paediatric practice and will be a useful experience to draw upon.

Aside from hands-on experience, getting involved in any research or audits taking place in the paediatric department is also a good idea. These can form the basis for departmental or regional presentations and could even be submitted to paediatric regional or national conferences. While in medical school, students should see if there is a paediatric society to join and become active members. If there isn't, they could perhaps establish one, with the aims of hosting paediatric teaching events at their university.

### CAREER OPTIONS WITHIN PAEDIATRICS

There are two types of clinical paediatric jobs: general paediatricians and sub-specialists (*Table 2*).

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