

A Framework of Competences

for Level 3 Training in Paediatric Inherited Metabolic Medicine

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CONTENTS

FOREWORD by Dr Patricia Hamilton.....	5
Section 1 Introduction	7
Progression in the Professional Development of a Paediatrician	9
What is a Paediatrician?.....	10
Contexts for Learning	11
Section 2 General Competences	13
Knowledge and Understanding	13
Skills.....	14
Values and Attitudes	17
Teaching and Research	18
Leadership and Management.....	19
Personal Commitment to Professional Standards.....	21
Communication Skills in Paediatrics.....	22
Section 3 General Clinical Competences	25
Development.....	25
Emotional development.....	25
Social development.....	26
Educational development.....	26
Growth and Nutrition	26
Adolescence.....	27
Section 4 Specialty-specific Competences in Paediatric Inherited Metabolic Medicine	29
Disorders of aminoacid and peptide metabolism (Phenylketonuria including the management in pregnancy, homocystinuria, MSUD, etc).....	30
Disorders of organic acid metabolism (propionic acidaemia, methylmalonic acidaemia, etc)	31
Hyperammonaemia and urea cycle disorders	31

Disorders of carbohydrate metabolism (Glycogen storage disease, galactosaemia, fructosaemia, etc)	31
Disorders of fatty acid oxidation (MCAD, LCHAD, etc).....	31
Disorders of ketone body metabolism	32
Lysosomal storage disorders (MPS, etc).....	32
Disorders of lipoproteins and lipid metabolism	32
Peroxisomal disorders (Adrenoleucodystrophy, Zellweger's syndrome, etc)	32
Disorders of purine and pyrimidine metabolism	32
Disorders of metal metabolism (Wilson's disease, Menkes syndrome, etc)	32
Congenital lactic acidoses and metabolic myopathies	32
Disorders of vitamin metabolism (biotin, cobalamin, etc)	32
Defects of membrane transport (lysinuric protein intolerance, etc)	32
Defects of glycosylation.....	32
Disorders of neurotransmission	33
Section 5	34
Practical Procedures and Investigations	34
Diagnostic Procedures	35
Therapeutic Procedures	36
Pharmacology and Therapeutics.....	37
References	38

FOREWORD

I am very pleased to present this competency framework for the new sub-specialty of Inherited Metabolic Medicine. This adds to the preceding frameworks we have published for Level 3 training, which build on the competency frameworks for Levels 1 and 2. The emphasis is firmly on the achievement of competences and not on a time-based training programme. From August 2007 all trainees entered specialty training within the framework outlined in the Modernising Medical Careers initiative. Achievement of these competences will determine a trainee's progress through the specialty training programme.

The development of this Level 3 framework, together with our other sub-specialty curricula, represents a huge amount of work. I would like to thank all those who have shown their commitment and support to this project.

In particular, we would like to thank Dr John Walter for leading the work on behalf of the sub-specialty. We would also like to thank Dr Edward Wozniak and Dr Gabrielle Laing, Chairs of General Paediatrics and Community Child Health CSACs respectively, who have been involved in the development of the competences at all three levels. Also Dr Mary McGraw and Dr Ian Doughty, the College Officers responsible for training, who have guided and supported this work unstintingly for their commitment and involvement in the project. In addition, we would like to thank Kim Brown for the initial development and co-ordination of this work.

These documents have been prepared for submission to PMETB in February 2008 and we hope to publish them, subject to their approval, by summer 2008.

Patricia Hamilton
President, Royal College of Paediatrics and Child Health

Section 1 Introduction

Who is this book for?

It is for doctors at Level 3 in their training in Paediatric Inherited Metabolic Medicine, their tutors and educational supervisors.

Why do I need it?

The book gives you and your tutors guidance about the areas you need to cover during your training. It gives a clear picture of what you have to have achieved by the end of this stage of training, before you become a consultant. You need this book as it forms the basis of your assessment at the end of Level 3 Training.

How do I use the book?

You can sit down with the book on your own and use it to help you identify areas of practice that you need to work on and those areas in which you feel fairly confident. You can talk to your tutor about the balance of your experiences and look for ways to ensure that you cover all the areas you need to.

Progression

This is the final stage in your training as a paediatrician. The competences you gained during Level 1 (Basic Specialist Training) and Level 2, Core Higher Specialist Training have formed the basis for your progression into Level 3 training and on to a Consultant post. Table 1 (page 9) illustrates this progression through your training.

A note about the format of this document

This framework sets out the competences that you need to achieve by the end of Level 3 Training. These build on and develop statements of competence set out for Levels 2 and 3. You are expected to work from all three documents throughout this final stage of your training to ensure that you maintain and continue to develop areas of competence already acquired as well as developing new ones.

Sections 2 and 3 present new statements of competence for Level 3 only, in order to keep the focus clear. Trainees will need to refer back to previous documents for Level 1 and 2 competences in General and General Clinical competences

A note about assessment

The statements in this book have been expressed as *learning objectives*. These are the focus of your training.

When it comes to your assessment, at the end of this phase of your training, we will want to know how *well* you have achieved these objectives and to be confident that you are fit to practise as a Paediatric Consultant. This is what we mean when we talk about your *competence*. So while here you may have, for example, a number of detailed objectives relating to consultation skills or communicating with children, in your assessment we will want to see how you bring all these together and how competent you are overall in your communication skills. This document is not intended as an assessment document but to support training. The assessment of your competence will be by work-based assessments already in use and currently being developed.

Working group:

Kim Brown	Training and Assessment Adviser
John Walter	Chair, Specialist Advisory Committee for Inherited Metabolic
Medicine	
Ian Doughty	Officer for Specialist Training
Mary McGraw	Vice-President for Training and Assessment

Progression in the Professional Development of a Paediatrician

During Level 1	During Levels 2 and 3	Continuing development as a consultant
<i>Acquires fundamental knowledge base</i>	<i>Applies knowledge base to provide appropriate clinical care</i>	<i>Evaluates knowledge and modifies clinical care pathways to enhance patient care</i>
<i>Acquires clinical examination and assessment skills and applies these in clinical practice</i>	<i>Analyses clinical findings to derive appropriate differential diagnosis and management plans</i>	<i>Evaluates assessment findings; refines and modifies management plans</i>
<i>Acquires all basic technical skills and basic life support</i>	<i>Proficient at all basic technical procedures, some complex procedures and provides advanced life support</i>	<i>May relinquish some skills in these areas dependent on area of clinical practice. May acquire speciality specific skills</i>
<i>Performs allocated tasks and begins to plan tasks</i>	<i>Plans and prioritises tasks appropriately</i>	<i>Increasing expertise with evaluation of priorities and appropriate delegation across a wide range of professionals</i>
<i>Performs allotted teaching tasks</i>	<i>Plans and delivers teaching to trainees and other professionals. Develops peer mentoring skills</i>	<i>Plans and modifies curricula. Performs assessment and appraisal. Able to provide mentorship</i>
<i>Aware of management issues</i>	<i>Develops management skills and able to take responsibility for a defined project. Contributes to Committees</i>	<i>Can negotiate and deal with conflict. Can contribute to and lead committees. Evaluates and modifies management structures</i>
<i>Performs allocated audit projects and understands the audit cycle</i>	<i>Designs audit project and understands risk management. Able to write appropriate clinical guidelines. Understands the Clinical Governance implications</i>	<i>Facilitates audit, and evaluates results. Evaluates guidelines and ensures implementation of appropriate changes</i>
<i>Understands the principles of critical appraisal and research methodology</i>	<i>Able to appraise the literature critically and apply to clinical practice</i>	<i>Able to evaluate critical appraisal performed by others. Able to lead research projects and support others in research</i>
<i>Works in multi-professional team</i>	<i>Able to take the lead and accept leadership from other members of the multi-disciplinary team</i>	<i>Evaluates and modifies multi-professional team-working</i>

What is a Paediatrician?

Paediatricians have a detailed knowledge and understanding of diseases in children. They are skilled in looking at health and ill-health in babies, children and adolescents, and at specific health issues, diseases and disorders related to these stages of growth and development. They develop expertise in practical procedures specifically related to the good clinical care of small babies and children. Paediatricians work in multi-disciplinary teams and with colleagues from a wide range of professional groups in hospitals, general practice and in the community, in social services and schools and with the voluntary sector. They have strong communication and interpersonal skills and take on a variety of roles within their different communities of practice. They share expertise effectively and assume the responsibilities of teaching, leadership and management roles where appropriate. They work with colleagues to ensure consistency and continuity in the treatment and care of children and young people in all aspects of their well-being. They are committed to a policy of advocacy for a healthy lifestyle in children and young people and for the protection of their rights.

Paediatricians are doctors who have a particular compassion and respect for children, young people and their families and enjoy working with them. They have an expert understanding of the ways in which illness affects the child, the parents and the rest of the family and are skilled in the management of emotionally complex family situations. They show patience and sensitivity in their communications with children and their families and a particular ability to explore each individual's perspectives of a problem. They are aware of religious and cultural beliefs that parents might hold about the treatment of their children. They know how to respond in these cases, when to seek support and where to find legal and ethical guidelines to support their practice.

Paediatricians ensure that they are up-to-date in their practice and endeavour to promote evidence-based medicine where possible. They are keen to develop innovative approaches to teaching in paediatrics and to research. They are committed to the highest standards of care and of ethical and professional behaviour within their specialty and within the medical profession as a whole. Central to their work is the principle that all decisions should be made in the best interests of the child or young person in their care.

Contexts for Learning

In drawing up this framework of competences, we have envisaged a wide range of opportunities in which trainees will learn. They will be expected to work on their own, using databases and electronic libraries to research particular conditions or areas of professional practice. This work might be in preparation for a clinic, or a presentation to a group of trainees and supervisors. Trainees may engage with a distance-learning programme in order to develop greater expertise in an area that interests them or that they need to strengthen. Supervisors will need to ensure opportunities for trainees who have undertaken independent study of this kind to share their learning with others. It is through teaching about something you have read or learned or understood differently that learning is consolidated and questioned.

Feedback is essential at all stages of the teaching and learning process. Even in the course of a lecture or on a ward round trainees can communicate important messages about their learning needs through facial expressions and body language as well as in their answers to questions. Tutors need to be ready to pick up on these, to seek out actively trainees' responses to their teaching so that they can make adjustments accordingly and ensure that effective learning takes place.

As well as independent study, trainees will find themselves in a number of different communities of practice. In many cases, their learning will result from shared discussion around the diagnosis of a condition, for example, or the identification of an injury. Experienced colleagues from a range of disciplines may join a specialist team, each sharing their expertise, in order to come to a safe diagnosis or decision about how to proceed, for example in the case of suspected abuse. Trainees need to be encouraged to join these discussions so that they develop confidence in their communication skills with colleagues and in their ability to contribute to clinical decision-making.

Supervisors need to be aware of the importance of asking questions in these situations and of the most effective way to do this, so that trainees are fully engaged and learning as they listen. Closed and open questions have their place, and explicit educational strategies such as 'scaffolding' are essential. The teacher takes the learners, step by step, from their initial level of understanding of a condition or a process, for example, to a deeper or more extensive understanding, through a formal cycle of informing, questioning, informing, testing out and consolidating new information. Ward rounds and clinics offer good opportunities for this kind of

interaction, with individual trainees and groups. The exchange may be brief or sustained and it is the close focus on understanding which gives this teaching technique its power. Trainees are also encouraged to write a reflective log and it is important to make clear that this needs to go beyond a narrative of events to an analysis of the process of learning they are experiencing. This, in turn, will equip them well to become teachers themselves of less experienced colleagues.

The college is committed to an explicit educational approach. Identifying contexts for learning is the first step. But it is important to go beyond this to a consideration of the way in which different contexts influence the teaching and learning that take place or that are required. It is helpful to be able to identify apprenticeship models, or experiential learning but these alone do not tell us much about the teaching and learning process. The curriculum sets out *what* is to be learned and we know *where* these things will be learned. But it is also essential to understand *how* teachers and trainees will learn.

With the completion of the curriculum, the college is now working on resources to support the teaching and learning of its contents and on guidance for trainees and supervisors on the pedagogical process of training in Paediatrics.

For an overview of teaching and learning in postgraduate medicine, see *Liberating Learning* (COPMED, 2002).

Section 2 General Competences

Knowledge and Understanding

Substantial re-wording or new statements of competence for Level 3 Training

- understand the impact of physical illness on mental functioning, for both children, young people and their parents and the effect of each upon the behaviour and functioning of the other
- understand the impact of relations and mental health upon a child's or young person's current and past emotions and behaviour
- understand the impact of culture and ethnicity in presentations of physical and psychological conditions
- know, understand and be able to compare and contrast medical and social models of disability
- understand the relationship between local health, educational and social service provision
- know about the agencies, both statutory and voluntary, that can provide general and condition-specific support to children, adolescents and their families in coping with their health problems
- know the objectives of paediatric follow-up
- understand and take account in their practice of risk issues to themselves and others, including those related to personal interactions, and bio-hazards
- have a working knowledge of risk assessment and its application to personal, professional, clinical and organizational practice
- understand and take account in their practice of measures to reduce clinical risk
- know how relative and absolute risks are derived and the meaning of the terms predictive value, sensitivity and specificity in relation to diagnostic tests
- know the legal and ethical guidelines to support their work, management and challenges and where to find more information when required
- be aware of the multidisciplinary investigation of sudden unexpected death in infancy
- understand the management of bereavement and be aware of national guidance documents on this
- understand the purpose of post-mortem examinations and know about procedures
- understand the process of bereavement in children and families and recognise abnormal grieving patterns

Competences specific to the specialty

By the end of Level 3 Training in Paediatric Inherited Metabolic Medicine, trainees will:

- know the normal physiology and biochemistry, including changes during childhood, relevant to metabolic disease
- know the principles of ex vivo and in vivo gene transfer and its relevance to Inherited Metabolic Medicine
- know the mechanisms of inheritance and understand molecular genetics, including mitochondrial DNA
- understand the principles of pre-natal diagnosis

- know the pathological and biochemical changes, clinical symptoms, investigations and management of metabolic disorders of these pathways and organelles
- know the metabolic indications and the long-term follow-up of liver transplantation, haematopoietic stem cell transplantation and renal transplantation
- know the principles of detailed therapy including the consequences of changes in the intake of nutrients
- understand the consequences of under-nutrition and specific nutritional deficiencies

- understand the impact on children and young people and their families of progressive metabolic disorders and those with a course which is difficult to predict

- have an understanding and experience of the drugs used for the treatment of metabolic disorders

(see Good Medical Practice (GMC 2006) - Good Clinical Care: 2, 3; Delegation and Referral: 45, 46.)

Skills

Substantial re-wording or new statements of competence for Level 3 Training

- recognise the breadth of different presentations of common disorders
- recognise the diseases and host characteristics which make certain presentations life-threatening and manage these situations with vigilance and appropriate urgency
- be able to recognise when both physical and psychological problems are present and when more than one condition or disorder may be present
- be able to assess and manage co-morbidities associated with the range of paediatric presentations

- take a history from a child, young person and parent of the presenting difficulties to acquire information in sufficient breadth and depth in a range of possible symptom areas to allow accurate formulation of the problem
- be able to undertake an assessment of the mental state of children and young people, taking into account their age and stage of development and know whether they have the skills to help them and when to seek more expert paediatric, mental health or psychiatric assessment
- have developed observation skills to support their interpretation of children's or young people's developmental levels and possible physical signs when they are unable to co-operate with formal assessments
- be able to supplement clinical assessment with standardised instruments or questionnaires
- know when to gather information from other professionals eg those working in education, social work or from others who see the child in a variety of settings
- be able to seek the views of children and young people, whatever their illness, regarding individual care and service planning, using expert resources appropriately
- be able to make a decision on the 'most likely' diagnosis and discuss this effectively with children and young people and their parents or carers, and with other colleagues, in the context of a plan of investigation and management
- be able to formulate a management plan for complex cases
- be able to review and modify a management plan as appropriate and know when to request help from senior colleagues or other services
- be able to take responsibility for the longer-term management of common acute and chronic cases leading or working with the multi-disciplinary and multi-agency teams, sub-specialists or networks as appropriate
- have developed expertise in practical procedures specifically related to the clinical care of small babies and children, and young people
- be able to develop and work within care pathways
- be able to manage and know how to obtain support for the consequences of chronic illness for a child, young person and their family
- be able to work effectively in multi-disciplinary teams and with colleagues from a wide range of professional groups
- be able to interact effectively with professionals in other disciplines and agencies and from the voluntary sector
- be aware of their role in the team and of their impact in the team

- have developed skills in recording consultations accurately and sensitively whilst maintaining a good rapport with the young person and family
- have developed a wide range of effective age-appropriate communication skills specific to their work with babies, children, young people and their families
- have developed credibility in their relationships with children, young people and their families, and with colleagues through their knowledge and skills and experience in clinical practice and in their ability to work independently
- have developed strategies to manage a child's or young person's anxiety and personal anxieties
- have developed basic behavioural management skills with parents, children and young people and with other professional colleagues
- be able to recognise, acknowledge and manage different levels of parental anxiety
- be able to assess patterns of relationships and functioning within a family and how these might impact on a child's or young person's illness, seeking professional advice where appropriate
- have developed effective skills in the management of emotionally complex family situations
- be able to recognise indicators of stress or mental health problems in family members and communicate appropriately with relevant professionals
- be able to remain calm in stressful or high-pressure situations and take a timely, rational approach to the problem
- be able to approach new situations which require good clinical judgement with an analytic and informed approach
- show confidence and independence in decision-making in the care of patients
- be able to apply effectively to their practice the knowledge and understanding acquired during training
- have developed a reflective approach to their practice, with an awareness of their level of expertise and limitations and their development needs
- show an ability to learn from their previous good practice, and from clinical errors
- be able to practise evidence-based medicine and understand and analyse critically its limits
- be able to understand the limitations of guidelines, how to use guidelines effectively and when it is appropriate to work outside guidelines
- have skills in managing perceptions of presenting complaints and illness
- have developed skills in maintaining appropriate confidentiality in relation to the social situation of the child
- be able to discuss an assessment of the psychosocial health of a child or young person with the multi-disciplinary team while respecting patient confidentiality

Competences specific to the specialty

By the end of Level 3 Training in Inherited Metabolic Medicine, trainees will:

- be able to offer genetic counselling and counselling prior to pre-natal diagnosis

(See Good Medical Practice (GMC, 2006) - Good Clinical Care: 2,3; Maintaining Trust: 19; Working with Colleagues 34, 36; Probity: 50.)

Values and Attitudes

Substantial re-wording or new statements of competence for Level 3 Training

- be committed to a policy of advocacy for a healthy lifestyle in children and young people and for the protection of their rights
- understand national and contribute to local initiatives aimed at reducing inequalities in child health and well-being
- practise with compassion and respect for children, young people and their families and act as a role model for others
- adopt an open-minded approach to equality and diversity in their practice
- be aware of the effects of social, cultural and religious context and conflict upon families
- understand the importance of cultural diversity and the difficulties where religious and cultural beliefs that parents might hold about the treatment of their children are in conflict with good medical practice and know when legal and ethical guidelines will support your management or view of the situation
- have developed strategies to manage relationships where health-care beliefs might cause conflict
- be able to advise patients appropriately on debates and controversies in health care
- be sensitive to the effects of stigma on children and families in relation to medical conditions
- be able to work effectively with children, young people and parents or carers, to agree and help them follow management plans
- be able to work effectively with young people who may have or may develop health care beliefs which are in conflict with those of parents or professionals, and know when legal and ethical guidelines will support your management or challenge of the situation
- be able to accept complex and difficult challenges
- show an understanding of the importance of ensuring a healthy balance between professional and domestic priorities

- have the willingness to acknowledge and reflect on the way in which they may, influenced by their earlier life experiences, have an impact on perceptions of and interactions with young people, their families and professionals

(See Good Medical Practice (GMC, 2001) - Good Medical Practice: 1; Good Clinical Care: 5; Maintaining Trust: 19; Working with Colleagues: 36.)

Teaching and Research

Substantial re-wording or new statements of competence for Level 3 Training

- have developed a range of effective teaching and learning skills in a range of clinical contexts
- be able to identify learning needs in a wide range of professionals and build on this in their teaching
- be able to elicit and act upon feedback on content and presentation of teaching
- take responsibility for the training, supervision and assessment of undergraduates and trainees and other professionals such as nurses, teachers and social workers in and outside the specialty
- have developed skills in the presentation of information relevant to their clinical practice for a range of audiences, including spoken presentations at meetings, written information for children and families and training materials for different groups of colleagues
- be able to lead departmental teaching programmes, including journal clubs
- be willing to accept mentoring as a positive contribution to their own professional development
- be willing to learn from others, to discuss cases openly and to seek advice as appropriate and as necessary
- be able to participate in teaching and research on topics within their specialty and in related areas
- conduct research with honesty and integrity, seeking ethical approval where appropriate and safeguarding the interests of patients
- demonstrate an understanding of 'good clinical practice' for all aspects of the conduct of clinical trials
- demonstrate an understanding of the role of ethics committees for clinical studies and the process of ethics applications
- understand the techniques used in epidemiological studies
- demonstrate an understanding of how to perform and interpret systematic reviews, how they differ from narrative reviews and understand the principles of meta-analysis

- understand the difference between population-based assessments and unit-based studies and be able to evaluate outcomes for epidemiological work
- be able to develop clinical guidelines, understand how they are produced nationally and how these should be used to guide their own practice
- be able to evaluate research effectively in paediatrics and child health

(See Good Medical Practice (GMC, 2001) - *Teaching and Training, appraising and assessing: 13, 14, 15, 16; Probity: 51.*)

Leadership and Management

Substantial re-wording or new statements of competence for Level 3 Training

- be able to provide specialist support to hospital- and community-based paediatric services including primary care
- be able to take on a leadership role in a multi-disciplinary team when appropriate, for example by representing the health needs of a child, young person and their family at a discharge meeting, and know when it may be inappropriate to do so
- be able to work effectively in multi-agency teams, for example, with social workers and teachers, and have developed an awareness of their own role within the team and of the skills and expertise of others
- be confident to make decisions within a team and be aware of their impact on other team members
- be able to advise the team providing advanced life support and to liaise effectively with anaesthetic and PICU staff
- demonstrate effective leadership skills in clinical situations, for example through their ability to organise, prioritise and delegate, and be able to help others to develop these skills
- have skills and strategies to manage conflict effectively
- have understanding and skills to be able to participate effectively in clinical and management meetings
- have developed effective administrative skills including ways to make best use of secretarial resources
- be able to handle enquiries from the press and other media effectively
- recognise their own working preferences and accept different approaches of colleagues
- know how to respond appropriately to health service targets and be able to participate in the development of services
- be able to work with stake-holders so that a client- or patient-centred service is created and sustained

- have gained an understanding of national and local regulatory bodies, particularly those involved in standards of professional behaviour, clinical practice and education, training and assessment
- understand the value and limitations of evidence-based medicine
- use principles of evaluation, audit, research and development in standard-setting and in improving quality
- demonstrate responsibility for ensuring reliability and accessibility of both themselves and others in their team
- have effective skills in ensuring the responsible approach of others in their team to health, stress and well-being

Competences specific to the specialty

By the end of Level 3 Training in Paediatric Inherited Metabolic Medicine, trainees will:

- have experience of the day-to-day running of a paediatric metabolic service, including the management of admissions policies within the medical directorate system
- have an understanding of the management skills required for the development and use of resources in the metabolic unit, including budget control, contracting, strategic planning and writing a business plan

(See Good Medical Practice (GMC, 2001) - Working with Colleagues: 34, 35, 36, 39, 42.)

Personal Commitment to Professional Standards

Substantial re-wording or new statements of competence for Level 3 Training

- understand the duty of all professionals working with children to report concerns about child protection issues to Social Services
- be able to contribute to the implementation of national and local health policy initiatives
- know and follow key legal and ethical guidelines relating to confidentiality, consent to treatment, the right to refuse treatment, continuing changes in the law and its interpretation and be aware of variability in Scotland, Wales and Northern Ireland
- know when in the interest of the child it may be necessary to break confidentiality
- be able to generate local and evaluate national clinical guidelines and protocols in paediatric practice and public health and recognise the individual patient's needs when using them
- participate and take responsibility for clinical governance activities, and encourage and support colleagues in their participation
- be able to carry out audit in a range of settings in partnership with all stakeholders in order to identify best practice
- know about and participate in clinical and research special interest groups relevant to their specialty
- know how to find, review and maintain relevant knowledge in their specialty in order to maintain their fitness to practice
- ensure that they are up-to-date in their practice and promote evidence-based medicine where possible
- be able to evaluate their own performance critically
- be open about sharing and reviewing their practice with others
- be aware of local processes for dealing with and learning from clinical errors and to be able to work within them

(See Good Medical Practice (GMC, 2001) - 1; *Maintaining Good Medical Practice*: 10, 12; *Relationships with Patients*: 17; *Working with Colleagues*: 35; *Dealing with Problems in Professional Practice*: 26, 27, 29, 30. *Probity*: 58.)

See also for all of these sections: *Good Medical Practice in Paediatrics and Child Health*,² London: Royal College of Paediatrics and Child Health (2002).

²*Good Medical Practice in Paediatrics and Child Health*, London: Royal College of Paediatrics and Child Health (2002). Online at www.rcpch.ac.uk/publications/recent_publications.html

Communication Skills in Paediatrics

Substantial re-wording or new statements of competence for Level 3 Training

- understand the importance of directing communications to the baby, child or young person as well as to parents and carers
- have developed skills to establish a child's or young person's and family's understanding of a situation from what has been said and written and to build on this effectively in discussion about the condition and its management
- understand the importance of seeking the views of all children and young people to inform decisions about their individual care and to encourage their participation in their care
- encourage children and young people to participate in their individual care and in the development of services, using expert resources appropriately
- have effective active listening skills in consultations with children and young people and understand the need to respect their views in accordance with their age and maturity and to respond appropriately where, for example, a child or young person is felt to be vulnerable
- have developed effective skills in working with children, young people and families to achieve concordance in planning management and treatment, enabling children and young people to maximise control over their illness and its management
- be able to respond appropriately, and know where to find assistance, in cases where a child, young person or family may not all speak English or where there is a sensory impairment that may affect understanding

- be able to respond to babies, disabled children or young people who may not be able to express themselves verbally, including those who might be in pain or distress
- be able to recognise, interpret correctly and respond to verbal and non-verbal cues from children, young people and parents
- have developed observation skills to support their interpretation of children's or young people's developmental levels and possible physical signs when they are unable to co-operate with formal assessments
- demonstrate appropriate responses and empathy for children, young people and their families experiencing difficulty and distress
- have developed a range of language strategies, such as the use of metaphor or images which relate to everyday life, to explain clearly to a child or young people and their family, their symptoms, condition or treatment, their feelings or behaviour

- be able to counsel parents about serious conditions and abnormalities within their area of expertise
- have effective strategies for careful and appropriate use of language in difficult and challenging circumstances, for example, at the birth of a baby with disabilities or where there is a conflict with colleagues
- be able to discuss the indications, benefits and adverse events of a procedure to patients, relatives and carers in a manner that will allow informed consent
- have developed a range of approaches to communicating the breadth of diagnostic possibilities and other clinical information to children, young people and their families so that consent is always informed and the plan and progress of treatment understood
- be able to advise children, young people and their families about the importance of concordance and about medication interactions and side-effects
- be able to convey and share effectively difficult or bad news, including end-of-life issues, with children, young people, parents or carers and help them to understand any choices they have or decisions to be made about ongoing management
- be able to prepare and discuss with parents, carers and other professionals “Do not attempt resuscitation” policies as appropriate, taking due account of the Human Rights Act (1998), ensuring that the best interests of the child are held as paramount at all times
- be able to seek consent for post-mortem examinations and communicate effectively with the Coroner
- be able to explain the role of other professionals and agencies to children, young people and their families
- have the confidence to be firm and diplomatic in difficult situations, for example, when dealing with angry parents
- understand the limits of their competence, particularly in stressful situations and be willing to seek help in managing sensitive and complex situations
- be able to demonstrate to trainees how to communicate a diagnosis and prognosis effectively to children, young people and their families
- be able to demonstrate and explain to trainees strategies used to conduct effective consultations with babies, young children, adolescents and their families
- have effective skills in written communications for a range of audiences, for patients and their families, colleagues and other professional organizations
- ensure that spoken and written communications with patients and families are presented in clear, straightforward English, avoiding jargon whenever possible
- ensure that written information in the form of booklets, leaflets, information sheets and websites support verbal communications wherever possible

- ensure that written communications summarise accurately discussions with children, young people and parents or carers, and, to avoid confusion and anxiety, do not include information that was not part of the original discussion
- be able to prepare a court report as a professional witness and develop the skills to present such material in court
- be able to write reports that explain the condition of a child or young person to non-health personnel working in the courts, social services or education
- know how to write reports about alleged abuse of children and young people for social services or the courts
- be able to use electronic communication media, taking into consideration the principles of confidentiality outlined in the Data Protection Act
- be able to liaise with parent support and self-help groups when necessary
- have developed effective professional networks to support clinical practice and other activities, including research, education and management

Competences specific to the specialty

By the end of Level 3 Training in Paediatric Inherited Metabolic Medicine, trainees will:

- have developed effective communication and interpersonal skills with babies, children, young people and their families and with colleagues from a range of professional contexts, including research and laboratory staff and managers

Section 3 General Clinical Competences

Development

Substantial re-wording or new statements of competence for Level 3 Training

- know the range of patterns of normal development from birth to adulthood
- know and understand the range of children's or young people's psychological and social development, including the normal range and what is outside it
- be able to identify when patterns of development are abnormal and where there may be a risk of abnormality which may only become apparent with time
- know the causes of disability, how disability might affect clinical examination and assessment and be able to contribute to a multi-disciplinary approach to management
- understand the severity of the presentation, taking into account normal development in appropriate domains
- understand the ways in which children's or young person's mental health difficulties may present in infancy, childhood and adolescence
- understand the impact of biological factors, including genetic and cognitive factors, on the mental health of children and young people
- understand the impact of other environmental factors (including violence, trauma, neglect, abuse and disruption, wherever this has occurred) on a child's development, mental health and functioning
- be able to assess the effects of recurrent or chronic illness and its treatment on growth, psycho-social, emotional, physical and sexual development and have strategies to minimize adverse effects

- know how to institute further assessment and investigation
- know about different modes of screening and health promotion strategies

Emotional development

Substantial re-wording or new statements of competence for Level 3 Training

- understand and recognise somatisation disorders and know how to provide initial management and how to access appropriate support
- recognise pointers to fabricated and induced illnesses and know how to provide initial management and how to access available support
- understand the emotional impact of illness and hospitalisation on children, young people and their families and take action to minimize this impact

- understand how a family's, child's or young person's attitude to the problem and services may have a significant impact on the presentation and its management
- recognise the need for specialised input in cases of serious emotional distress or mental illness and ensure their needs are met within local health provision
- understand the emotional dimensions of eating disorders and recognise and initiate treatment
- be able to assess parenting skills and recognise and respond to indications of unsatisfactory or unsafe parenting
- know how to access help in cases where children or young people of different ages might be deprived of opportunities to play and to learn
- know how to manage common behavioural problems

Social development

Substantial re-wording or new statements of competence for Level 3 Training

- be able to recognise and understand the impact of autistic spectrum disorders and other organic disorders on social development

Educational development

Substantial re-wording or new statements of competence for Level 3 Training

- demonstrate, in all aspects of their practice, an understanding, of the vulnerability of a child or young person with learning difficulties

Growth and Nutrition

Substantial re-wording or new statements of competence for Level 3 Training

- know the reasons for faltering growth, including emotional factors and how to investigate appropriately
- understand and assess normal and abnormal pubertal development and its relationship to growth
- understand the environmental factors contributing to obesity and how these might be altered
- be able to recognise feeding problems and work with parents directly to offer simple advice and to treat co-morbid conditions

- know about the principles and methods and indications for nutritional support and common problems that may arise from invasive methods or refeeding
- be able to identify nutritional deficiencies and growth failure which may occur in children and young people who undergo unsupervised dietary modification

Adolescence

Substantial re-wording or new statements of competence for Level 3 Training

- understand what the specific needs of young people are, in terms of their emotional, mental and physical health, and how these are different from those of children
- know the epidemiology of the main causes of morbidity and mortality in young people
- ensure that young people have access to ‘in-patient’, ‘outpatient’ and other medical services that best meet their needs
- understand why young people harm themselves and respond appropriately to actual or threatened episodes of self-harm in adolescents
- understand the consequences of self-harm and be able to work as part of a clinical network in the management of the young person who self-harms
- be able to discuss sexual health issues including basic contraceptive advice and know how to help the young person access appropriate sexual health or genetic advice
- know about national policies concerning the health care of young people, including those which help to reduce teenage pregnancy
- understand the processes of adolescence including experimental behaviours, learning by experience, achieving independence from the family, and the consequences of these on health and illness in young people
- be able to discuss comfortably with young people important health behaviours such as the use of tobacco, alcohol or recreational drugs, and intimacy and sexual activities together with the promotion of appropriate strategies for these in relation to specific conditions such as asthma, diabetes, cystic fibrosis, physical disability
- understand the particular needs of adolescents with regard to their independence and autonomy, education and work, body image and sexual identity, concordance with medication and risk-taking and understand how these factors may be affected in young people with chronic conditions

- be able to support young people in self-management of both acute and chronic disease where they want to, and have an understanding as to how to best help when the young person cannot or does not want to manage this
- be able to discuss the implications of chronic illness or disability for career options where appropriate and at a negotiated time, be able to raise and agree management of end-of-life issues with young people and their families and record conclusions in medical notes
- understand issues around transition from paediatric to adult care in adolescents with chronic conditions and disabilities, and be able contribute effectively to transitional care services
- understand and value the roles of members of the multidisciplinary team in the delivery of a transitional care programme

Section 4 **Specialty-specific Competences in Paediatric Inherited Metabolic Medicine**

By the end of Level 3 Training in Paediatric Inherited Metabolic Medicine, trainees will:

- have a detailed understanding of biochemical pathways and cellular organelles that can be affected by inherited metabolic diseases
- have a good working knowledge of genetics particularly as applicable to inherited disease
- know the clinical presentation of the range of disorders in inherited metabolic disease, metabolic disturbances, inheritance, diagnostic tests, treatment and prognosis
- be able to recognise the dysmorphology associated with certain inherited metabolic disorders
- be able to apply detailed knowledge of the various clinical presentations of inherited metabolic disorders to forming a differential diagnosis, and arranging appropriate investigations and management
- know the criteria and methodology for newborn screening for inherited metabolic disease
- have a detailed knowledge of which tests are indicated for the common and complex inherited metabolic disorders
- understand the principles and methods of investigations and be able to interpret results effectively
- know which investigations should be used for the monitoring of long-term disorders
- be able to interpret investigations accurately and know the analytical, physiological and nutritional factors that can influence the results
- know the indications for and be able to plan relevant tissue biopsies and be able to interpret the results
- be able to perform skin biopsies for the investigation of inherited metabolic disorders
- know when molecular genetic investigations are indicated and be able to interpret the results
- know the methods available for prenatal diagnosis for inherited metabolic disease and be able to counsel parents appropriately
- be able to interpret the results of newborn screening and know the actions that need to be taken on a positive screening result
- know the implications of a positive screening and be able to talk to parents about positive screening tests
- understand the principles of therapy for inherited metabolic disease, including reducing the load on affected pathways, correcting product deficiency, decreasing metabolic

toxicity, stimulating residual enzyme activity, transplantation, pharmacological enzyme replacement, and gene therapy

- have a detailed knowledge of the pharmacological agents used in the treatment of inherited metabolic disease
- be able to work effectively with specialist colleagues around dietary management, including assessment of nutritional intake and requirements, the use of specific diets for particular conditions, in long-term and emergency situations
- know when to consult other sub-specialists from other disciplines and be able to work with them in the management of patients
- know the indications for patients to require paediatric intensive care support including assisted ventilation
- know the indications for extracorporeal therapy
- be able to give clear, appropriate and accurate written or phone advice to general paediatricians and other colleagues when consulted about the diagnosis or on-going management of patients
- be able to use on-line databases, including OMIM and other on-line resources, to obtain up-to-date information on particular inherited metabolic disorders
- be able to work closely with metabolic dieticians, clinical biochemists and other members of the multidisciplinary team in the management of patients with inherited metabolic disease
- be able to formulate written protocols for the diagnosis and management of inherited metabolic disease
- be able to liaise appropriately with the parents' of children with inherited metabolic disease and patient support groups

Inborn errors of metabolism

The competences set out below indicate the levels of knowledge and skills required for Level 3 training for a range of inherited metabolic disorders.

Disorders of aminoacid and peptide metabolism

- be able to take responsibility for the management of individuals with phenylketonuria (including management in pregnancy)
- understand the dietetic treatment of this condition and to work closely with the metabolic dietician in order to achieve optimum treatment
- know how to investigate for other causes of hyperphenylalaninaemia and to manage children with bipterin defects

- be able to manage other disorders of aminoacid metabolism including homocystinuria, maple syrup urine disease and tyrosinaemia
- know about the various dietetic products available for the treatment of aminoacid disorders and how these should be used

Disorders of organic acid metabolism

- know the range of clinical presentations that can occur in propionic academia, methylmalonic acidaemia and other organic acid disorders
- be able to ensure adequate nutrition in these group of disorders
- know their long term complications and the arguments for and against the use of organ transplant in these conditions
- know how to investigate for vitamin responsive organic acids disorders

Hyperammonaemia and urea cycle disorders

- know when to suspect hyperammonaemia
- know the methods of measuring blood ammonia and how the results may be affected by sampling and sample processing
- know the causes of hyperammonaemia and be able to investigate appropriately in order to establish the underlying diagnosis
- be able to manage acute hyperammonaemia and to arrange extracorporeal treatment if required
- be able to undertake the long term management of urea cycle disorders

Disorders of carbohydrate metabolism and gluconeogenesis

- be able to undertake the appropriate investigation for the glycogen storage disorders and to provide effective management
- know the presentation of the galactosaemias and their management.
- advise parents on the long term problems associated with classical galactosaemia
- know the presentation, method of investigation and management of other disorders of carbohydrate metabolism including fructosaemia and those of glucose transport
- know the presentation, method of investigation and management of disorders of gluconeogenesis including fructose-1,6-bisphosphatase deficiency

Disorders of fatty acid oxidation

- know the possible clinical presentations of disorders of fatty acid oxidation
- be able to interpret blood acylcarnitine profiles from tandem mass spectrometry
- know the other investigations necessary in order to establish a diagnosis

- have a detailed knowledge of the newborn screening programme for MCAD deficiency and the action required following a positive screening result both in terms of confirming the diagnosis and immediate action required
- manage patients with MCAD deficiency diagnosed from newborn screening or following clinical illness
- have a detailed knowledge investigation and management of the rarer fatty acid oxidation disorders including LCHAD deficiency

Disorders of ketone body metabolism

- know how disorders of ketone production present, the difficulties that may be encountered in their diagnosis and how they should be managed

Lysosomal storage disorders

- have a detailed knowledge of the clinical presentation of the mucopolysaccharidoses, oligosaccharidoses and disorders of sphingolipid metabolism .
- know the investigations necessary to establish the correct diagnosis
- know which patients may benefit from enzyme replacement therapy and to know how such treatment can be arranged
- know the indications for haematopoietic stem cell transfer

Disorders of lipoproteins and lipid metabolism

- be able to manage patients with hyperlipidaemia, including those with familial homozygous hypercholesterolaemia
- know the dysmorphology and other clinical features associated with disorders of cholesterol synthesis

Peroxisomal disorders

- be able to investigate children with suspected disorders of peroxisomal metabolism and be able to undertake appropriate management where the diagnosis is confirmed.
- manage patients with X-linked adrenoleucodystrophy and, in particular, know how asymptomatic patients should be monitored

Disorders of purine and pyrimidine metabolism

- know the catabolic and salvage pathways of purine and pyrimidine metabolism, the major presenting features of disorders of these pathways and the laboratory findings
- be able to undertake the management of patients with a confirmed diagnosis

Disorders of metal metabolism

- know the different clinical presentations of Wilson disease and the appropriate pharmacological agents to treat this condition
- be able to recognise and diagnose Menke disease

Congenital lactic acidoses and metabolic encephalomyopathies

- have a detailed knowledge of mitochondrial oxidative phosphorylation and the genetics of mitochondrial cytopathies
- be aware of the wide range of possible clinical presentations
- be able to manage the investigations for suspected disorders
- be aware of the difficulties of making a diagnosis and providing genetic counselling, including prenatal diagnosis

Disorders of vitamin metabolism

- have a detailed knowledge of disorders of vitamin metabolism, their clinical presentation, the investigations necessary to confirm vitamin responsiveness and their management

Defects of membrane transport

- know the clinical presentations of this groups of disorders, their inheritance, diagnostic investigations and management

Defects of glycosylation

- know the clinical presentation and diagnostic protocol for the diagnosis of the congenital disorders of glycosylation

Disorders of Neurotransmission

- know the metabolic pathways involved in neurotransmission and the disorders associated with these including inborn errors of gamma aminobutyric acid and of receptors and transporters of neurotransmitters.
- know the methods necessary to investigate these disorders and their management

Section 5 Practical Procedures and Investigations

By the end of Level 3 Training, trainees will:

- know the appropriate indications for practical procedures and investigations
- know the contraindications and complications of procedures
- know the local and national guidelines for obtaining informed consent
- obtain informed consent appropriately – delete? or add to above
- know the local and national guidelines for undertaking investigations or procedures
- know the local guidelines for providing sedation and pain relief for practical procedures
- know the relevant anatomical markers for invasive procedures
- know and practise scrupulous aseptic techniques
- have developed confidence in independent performance of practical procedures
- be able to supervise and teach others
- recognise complications of procedures and be able to respond appropriately
- understand and follow the local guidelines for the prevention and management of needle-stick injury
- be able to recognise the importance of universal precautions as well as the disposal of sharps within the department
- know about processes for critical incident reporting
- be aware of safety issues for patients and staff in relation to investigations of body fluids and radiation
- understand the importance of post-mortem investigations
- know the national and local guidance for obtaining consent for post-mortem
- be able to interpret results of investigations requested and respond appropriately
- be able to give appropriate medical information when requesting investigations
- know that results should be requested clearly and retrieved promptly
- understand common age-appropriate normal ranges or appearances
- be able to use all equipment required to undertake common procedures and investigations
- know about the role of complex investigations eg CT and MRI scans and their diagnostic potential and complications
- be able to record results and document procedures legibly and accurately
- be aware of the factors that are likely to influence the anxiety of the child, parent and doctor and know how to enlist effectively the help of play-leaders, nursing staff and more senior paediatric staff when necessary
- be receptive to feedback from patients and parents/carers on the effects of medication/treatment

- be able to explain the investigation results to parents and/or the child
- have experience of speaking to parents when complications have occurred

- recognise when the results of commonly-used radiological investigations are abnormal
- supervise handover of results that still need to be obtained at the end of shifts

Competences specific to the specialty

By the end of Level 3 Training in Paediatric Inherited Metabolic Medicine, trainees will:

- know the indications for tests, biopsies and screening methods for a range of metabolic disorders and be able to plan and interpret these accurately
- understand the use of investigations for the long-term monitoring of metabolic disorders
- be able to interpret investigations, taking into account analytical, physiological and nutritional factors which might influence the results
- have experience of the use and interpretation of molecular genetic techniques
- developed skills in practical aspects of dietary management
- be able to interpret psychological and neuro-psychometric tests

Diagnostic Procedures

By the end of Level 3 Training, trainees will be able to perform the following diagnostic procedures independently:

- collection of blood from central lines
- umbilical artery and venous cannulation and sampling
- peripheral arterial cannulation
- venepuncture
- capillary blood sampling
- suprapubic aspiration of urine
- urethral catheterisation
- routine testing of urine
- perform basic lung function tests
- electrocardiogram
- lumbar puncture
- non-invasive blood pressure measurement

Competences specific to the specialty

By the end of Level 3 Training in Paediatric Inherited Metabolic Medicine, trainees will know the indications for, be able to perform, interpret and teach the following diagnostic procedures:

- controlled fasting test
- 24 hour glucose/lactate profile
- glucagon test
- tests for co-factor responsive disorders (vitamin B12; bipterin, pyridoxine)
- loading tests (glucose, galactose, fructose, protein, and allopurinol)
- exercise tests (semi-ischaemic forearm exercise test, bicycle ergometric test, treadmill test)
- skin biopsy for fibroblast culture
- collection of CSF for neurotransmitters
- postmortem protocol

They will have experience of the use and interpretation of:

- molecular genetic tests

and they will be able to interpret:

- psychological and neuro-psychometric tests

Therapeutic Procedures

By the end of Level 3 Training, trainees will be able to perform the following therapeutic procedures independently:

- administer intradermal, subcutaneous, intramuscular, intravenous injections
- percutaneous long-line insertion
- bag, valve and mask ventilation
- needle thoracocentesis for pleural effusion or pneumothorax
- tracheal intubation
- intubation of newborn infants of most gestations
- administration of surfactant
- external chest compression
- insertion of intraosseous needle

Pharmacology and Therapeutics

By the end of Level 3 Training, trainees will:

- know and understand the pharmacological basis for treatments
- know the approved indications and justification for prescribing drugs in common paediatric problems
- know the pharmacokinetics and pharmacodynamics of commonly prescribed drugs
- be able to calculate drugs accurately according to specific dose for weight, or age/weight range or on a specific dose/surface area basis
- know the risks of prescribing in the child-bearing years, in pregnancy and in breast-feeding mothers
- know about the roles of the regulatory agencies involved in drug use, monitoring and licensing (for example the National Institute of Clinical Excellence, the Committee on Safety of Medicines, the Medicines and Healthcare products Regulatory Agency and Hospital Formulary Committees)
- be able to find out information necessary for safe prescribing through use of paediatric formularies and pharmacy liaison
- know about drug interactions of commonly used drugs
- know about procedures for obtaining consent in children and young people for the administration of drugs
- be able to use the local and national guidelines for the relief of pain in children
- know and follow local policies for intrathecal cytotoxic therapy
- respond appropriately to errors of prescription or administration and be able to talk to parents about this
- be able to prescribe safely and supervise prescription for the newborn, and for children of all ages
- know about the licensing of medicines for paediatric patients and unlicensed and off-label use and the implications of extemporaneous products
- know how to explain relevant potential adverse side-effects
- be able to advise and supervise safe prescription of intravenous fluids to medical and surgical patients
- be able to prescribe in a manner that enhances adherence and provide information and explanation that enhances concordance

Competences specific to the specialty

By the end of Level 3 Training in Paediatric Inherited Metabolic Medicine, trainees will :

- have a detailed knowledge of the medicines used for the treatment of inherited metabolic disease

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