

Using the care pathway

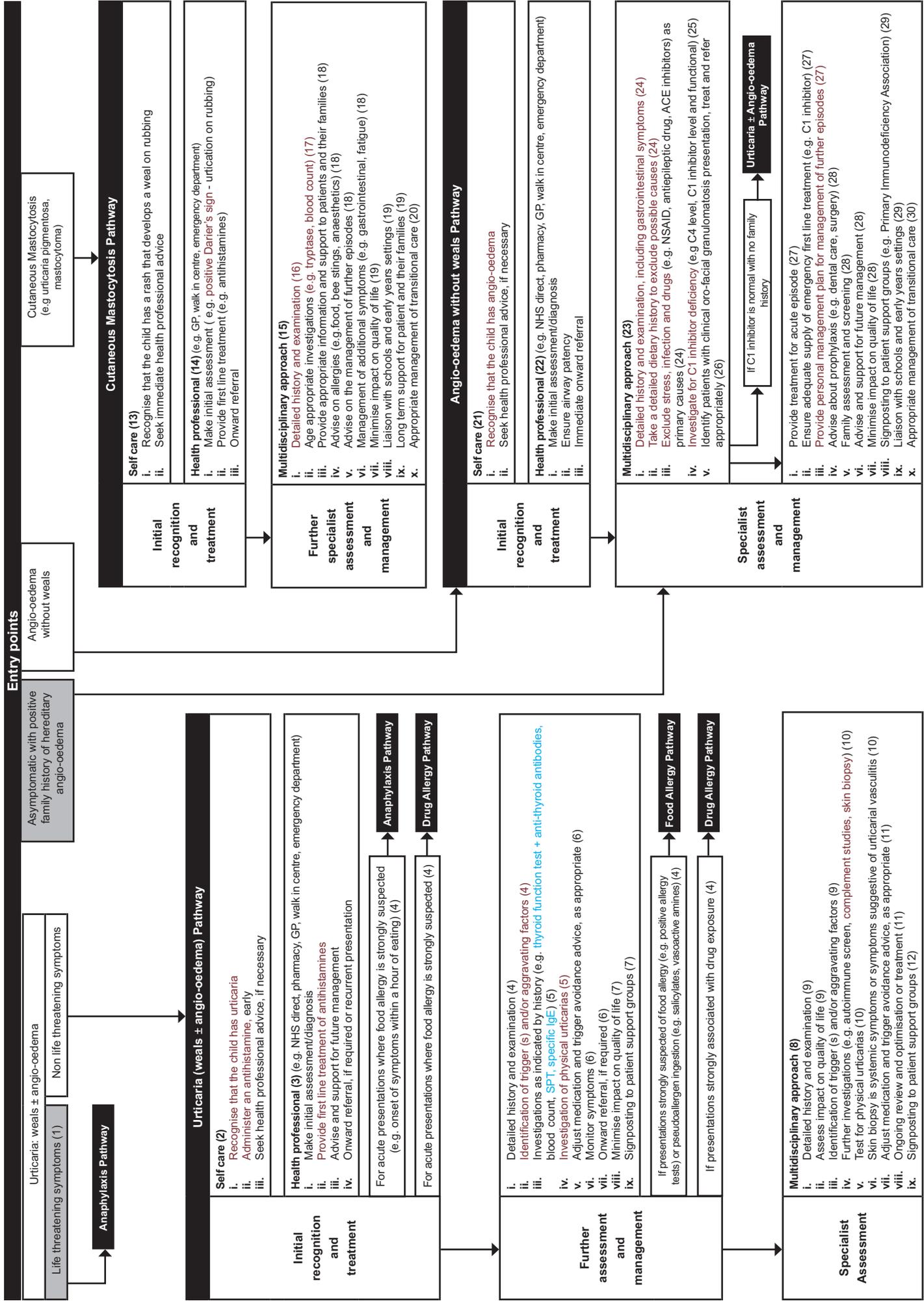
The Royal College of Paediatrics and Child Health (RCPCH) care pathway for urticaria, angio-oedema or mastocytosis is presented in two parts: an algorithm with the stages of ideal care and a set of competences required to diagnose, treat and optimally manage children with a urticaria, angio-oedema or mastocytosis. The algorithm has numbers which correspond to the competences outlined within the body of the document. These competences have not been assigned to specific health professionals or settings in order to encourage flexibility in service delivery. Each pathway has a set of core knowledge documents of which health professionals should be aware. These documents are the key clinical guidance that inform the pathways.

We recommend that this pathway is implemented locally by a multidisciplinary team with a focus on creating networks between staff in primary and community health care, social care, education and hospital based practice to improve services for children with allergic conditions. All specialists should have paediatric training in line with the principles outlined in the Department of Health Children's National Service Framework - particularly standard 3 which states that staff training should reflect the common core of skills, knowledge and competences that apply to staff who work with children and young people.

For the purposes of the RCPCH care pathways children is an inclusive term that refers to children and young people between the ages of 0-18 years. It is important to recognise that, while the RCPCH urticaria, angio-oedema or mastocytosis pathway is linear, entry can occur at any part in the pathway.

Further information regarding the RCPCH allergy care pathways can be downloaded at: www.rcpch.ac.uk/allergy.





Notes:

- The colours on the pathway and competence table correspond to the modified Scottish Intercollegiate Guidelines Network (SIGN) grade: **GRADE A** **GRADE B** **GRADE C** **GRADE D** **GRADE E** **GRADE F** **GRADE G** **GRADE H** **GRADE I** **GRADE J** **GRADE K** **GRADE L** **GRADE M** **GRADE N** **GRADE O** **GRADE P** **GRADE Q** **GRADE R** **GRADE S** **GRADE T** **GRADE U** **GRADE V** **GRADE W** **GRADE X** **GRADE Y** **GRADE Z** **GRADE AA** **GRADE AB** **GRADE AC** **GRADE AD** **GRADE AE** **GRADE AF** **GRADE AG** **GRADE AH** **GRADE AI** **GRADE AJ** **GRADE AK** **GRADE AL** **GRADE AM** **GRADE AN** **GRADE AO** **GRADE AP** **GRADE AQ** **GRADE AR** **GRADE AS** **GRADE AT** **GRADE AU** **GRADE AV** **GRADE AW** **GRADE AX** **GRADE AY** **GRADE AZ** **GRADE BA** **GRADE BB** **GRADE BC** **GRADE BD** **GRADE BE** **GRADE BF** **GRADE BG** **GRADE BH** **GRADE BI** **GRADE BJ** **GRADE BK** **GRADE BL** **GRADE BM** **GRADE BN** **GRADE BO** **GRADE BP** **GRADE 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- The numbers on the pathway correspond to the competences required to provide care - these are on the following pages
- Links to the references can be found within the competence statements

Definitions

Urticaria

Chronic urticaria is relatively common with up to 3% of children being affected and acute urticaria occurs more commonly with between 4.5-15% of UK children (29).

Hereditary angio-oedema (HAE) is rare with no reported bias in different ethnic groups. It has an estimated population prevalence of 1 in 50,000. Mastocytosis is uncommon, figures for prevalence are unknown.(30).

This is important because accurate diagnosis leads to targeted treatment. **Urticaria** is characterised by fluctuating weals and/or angio-oedema. A **weal** consists of three typical features:

1. a central swelling of variable size, almost invariably surrounded by a reflex erythema
2. associated itching or, sometimes, burning sensation
3. a fleeting nature, with the skin returning to its normal appearance, usually within 1-24hours

Chronic urticaria is characterised by continuous symptoms that persist for more than 6 weeks. Episodic urticaria is characterised by the occurrence of repeated relapses of urticaria with intervals of at least a week between each relapse. Urticarial vasculitis is distinguished by palpable purpura and bruising or discoloration that persists after the weal has disappeared. The urticarial lesions persist for more than 24 hours and respond poorly to antihistamines. The commonest cause in childhood is Henoch-Schonlein purpura

Angio-oedema

Angio-oedema is characterised by:

1. a sudden, pronounced swelling of the lower dermis and subcutis
2. which is pale rather than pink and may be painful rather than itching
3. frequent involvement below mucous membranes
4. resolution that is slower than for weals and can take up to 72 hours

In hereditary angioedema (HAE) this may also involve symptoms of abdominal pain due to intestinal oedema causing obstruction.

Mastocytosis

Mastocytosis is a heterogeneous group of disorders characterised by abnormal growth and accumulation of mast cells (MC) in one or more organ systems. The diagnosis of cutaneous mastocytosis (CM) in children is based on typical clinical and histological skin lesions and absence of systemic involvement. Most children present with maculopapular cutaneous mastocytosis (urticaria pigmentosa, UP). Other less frequent forms of CM are diffuse cutaneous mastocytosis (DCM) and mastocytoma of skin (31).

Competences: Urticaria (weals ± angio-oedema)

Ref	Pathway stage	Competences: Urticaria (weals + angio-oedema) Pathway
1	Life threatening symptoms	<p>Know</p> <ul style="list-style-type: none"> to consider the anaphylaxis pathway basic life support <p>Be able to</p> <ul style="list-style-type: none"> recognise that the child has life threatening symptoms (e.g. difficulty breathing, floppy) call for an ambulance or advise the patient to present to the ED immediately
2	Initial recognition and treatment - self care	<p>Know</p> <ul style="list-style-type: none"> the signs and symptoms of an urticarial rash (32) <p>Be able to</p> <ul style="list-style-type: none"> recognise that the child has urticaria (32) administer an antihistamine (32-34) early in an adequate dose seek appropriate health professional advice, if necessary
3	Initial recognition - health professional/NHS direct	<p>Know</p> <ul style="list-style-type: none"> the signs and symptoms of an urticarial rash (32) the possible causes of an urticarial rash (35-37) that urticaria may be one of the presentations of food or drug allergy or may be associated with infection to consider the food allergy pathway, drug allergy pathway or the anaphylaxis pathway <p>Be able to</p> <ul style="list-style-type: none"> make an initial assessment and diagnosis recognise that the child has urticaria (32) administer an antihistamine (33, 38), early in an adequate dose recognise when food allergy is a possible cause of urticaria refer onwards if symptoms do not settle
4	Further assessment and management - detailed history and examination, trigger(s) and aggravation(s)	<p>Know</p> <ul style="list-style-type: none"> the causes and exacerbating factors of acute urticaria and angio-oedema <p>Be able to</p> <ul style="list-style-type: none"> take an allergy focused history and examination (e.g. investigate allergic co-morbidities) (39) identify possible precipitating causes e.g. cold, exercise, sunlight, food (36) and drugs (34, 36) identify possible aggravating factors e.g. heat, stress, viral infections (36)

Ref	Pathway stage	Competences: Urticaria (weals + angio-oedema) Pathway
5	Further assessment and management - investigations	<p>Have</p> <ul style="list-style-type: none"> access to appropriate facilities, practical skill and knowledge to undertake and interpret investigations including <ul style="list-style-type: none"> skin prick testing (SPT) (40) serum specific IgE testing (40) thyroid function tests and thyroid auto-antibodies (41) full blood count appropriate quality control through guidelines and standard operating procedures to ensure the clinical competence of staff conducting SPT access to an appropriately accredited laboratory for in-vitro tests <p>Know</p> <ul style="list-style-type: none"> the indications for performing further investigations (36) how to interpret the results in light of the clinical history <p>Be able to</p> <ul style="list-style-type: none"> perform investigations for physical urticarias (e.g. ice cube challenge test) (36)
6	Further assessment and management - adjust medication/ advice, monitor symptoms, onward referral	<p>Know</p> <ul style="list-style-type: none"> the stepwise management of urticaria as outlined in the EAACI/ GA2LEN/EDF guideline for the management of urticaria (32) the side effect profile of medication(s) <p>Be able to</p> <ul style="list-style-type: none"> recognise the side effects of medication and adjust accordingly monitor symptoms and response to treatment advise about avoidance of trigger(s) refer onwards if symptoms are not controlled or are ongoing (e.g. chronic urticaria)
7	Further assessment and management - quality of life, signposting	<p>Know</p> <ul style="list-style-type: none"> how urticaria may impact on different aspects of daily life, including schooling <p>Be able to</p> <ul style="list-style-type: none"> liaise with schools and early years settings to minimise impact on quality of life provide information to patients/parents/carers about the natural history of urticaria provide information for patients to access patient information e.g. British Association of Dermatologists (42)
8	Specialist assessment - multidisciplinary team	<p>Know</p> <p>that this care should be provided using a multidisciplinary approach (e.g. paediatric allergist, dermatologist, immunologist, nurse specialist(s), dietitian, pharmacist, psychological support)</p>

Ref	Pathway stage	Competences: Urticaria (weals + angio-oedema) Pathway
9	Specialist Assessment - history/ examination, impact on quality of life, trigger(s)/ aggravations	<p>Know</p> <ul style="list-style-type: none"> the causes and exacerbating factors of acute and chronic urticaria and angio-oedema that specialist assessment occurs in addition to further assessment and management that patients with chronic urticaria may have greater impairment of quality of life <p>Be able to</p> <ul style="list-style-type: none"> take an allergy focused history and examination identify possible precipitating causes (e.g. cold, drugs, exercise, sunlight, food) and aggravating factors (e.g. heat, stress, viral infections) assess the impact on quality of life
10	Specialist Assessment - extended diagnostic programme, test for physical urticarias	<p>Have</p> <ul style="list-style-type: none"> access to appropriate facilities, practical skill and knowledge to undertake and interpret investigations including <ul style="list-style-type: none"> autoimmune profile complement studies skin biopsy (36) <p>Know</p> <ul style="list-style-type: none"> the indications for performing extended diagnostic programme (36) the indications for performing skin biopsy how to interpret the results in light of the clinical history the implications of systemic urticarial vasculitis (e.g. nephritis, proteinuria) <p>Be able to</p> <ul style="list-style-type: none"> perform extended diagnostic programme (36) advise and supervise a pseudo-allergen free diet for 3 weeks perform investigations for physical urticarias (e.g. ice cube challenge test) (36) commence treatment in light of the history and investigations
11	Specialist Assessment - adjust medications/ avoidance advice, review and optimise treatment	<p>Know</p> <ul style="list-style-type: none"> the stepwise management of urticaria as outlined in the EAACI/ GA2LEN/EDF guideline for the management of urticaria (32) the side effect profile of medication the indications for second line treatment for urticaria (33, 43-46) <p>Be able to</p> <ul style="list-style-type: none"> prescribe and monitor immunomodulatory therapy for urticaria recognise the side effects of medication and adjust accordingly monitor symptoms and response to treatment advise about avoidance of trigger(s)

Ref	Pathway stage	Competences: Urticaria (weals + angio-oedema) Pathway
12	Specialist Assessment – signposting to patient support groups	Be able to <ul style="list-style-type: none"> <li data-bbox="574 212 1420 280">▪ provide information for patients to access patient information e.g. <u>British Association of Dermatologists</u> (42)

Competences: Cutaneous Mastocytosis Pathway

Ref	Pathway stage	Competences: Cutaneous Mastocytosis Pathway
13	Initial recognition -self care	<p>Know</p> <ul style="list-style-type: none"> that the rash needs to be seen by a health professional <p>Be able to</p> <ul style="list-style-type: none"> seek health professional advice
14	Initial recognition – health professional	<p>Know</p> <ul style="list-style-type: none"> the signs and symptoms of cutaneous mastocytosis (47) <p>Be able to</p> <ul style="list-style-type: none"> recognise the different manifestations of cutaneous mastocytosis assess for a positive Darier’s sign (31, 48) (a rash that develops a weal on rubbing) provide symptomatic relief (e.g. antihistamine) refer for further assessment
15	Further specialist assessment and management	<p>Know</p> <ul style="list-style-type: none"> that this care should be provided using a multidisciplinary approach (e.g. paediatric allergist, dermatologist, haematologist, nurse specialist(s), dietitian, pharmacist, psychological support)
16	Further specialist assessment and management – detailed history and examination	<p>Be able to</p> <ul style="list-style-type: none"> take a detailed clinical history and examination (48)
17	Further specialist assessment and management – age appropriate investigations	<p>Have access to</p> <ul style="list-style-type: none"> measurement of serum tryptase <p>Know</p> <ul style="list-style-type: none"> the importance of measuring mast cell tryptase (31) the normal range for serum tryptase (31) <p>Be able to</p> <ul style="list-style-type: none"> perform appropriate blood tests (e.g. tryptase, full blood count, chemistry, routine coagulation parameters) (31) request mast cell tryptase measurement and interpret the results

Ref	Pathway stage	Competences: Cutaneous Mastocytosis Pathway
18	Further specialist assessment and management - advice/information, management of further episodes, symptoms, co-morbidities	<p>Be able to</p> <ul style="list-style-type: none"> ▪ provide information to patients and their families about the natural history of mastocytosis ▪ advise about the risk of severe reactions following exposure to allergens (e.g. bee and wasp stings, foods) and drugs (e.g. general anaesthetics) ▪ advise about medication where symptoms persist (e.g. H1 antihistamines, H2 antihistamines) ▪ advise about the provision of adrenaline injectors and personal management plans, where appropriate ▪ advise on the management of further episodes ▪ identify when symptoms may be due to mastocytosis and advise about their management (49)
19	Further specialist assessment and management - minimise impact on quality of life, signposting, liaison with schools and early years settings, long term support	<p>Know</p> <ul style="list-style-type: none"> ▪ the natural history of cutaneous mastocytosis and possible complications <p>Be able to</p> <ul style="list-style-type: none"> ▪ recognise where mastocytosis may impact on quality of life ▪ advise and support children and their families ▪ provide information for patients to access patient support groups e.g. UK Mastocytosis Support Group (50), MastoKids (51), NHS Choices – Mastocytosis (52) ▪ provide regular review and support as necessary
20	Further specialist assessment and management - transitional care	<p>Know</p> <ul style="list-style-type: none"> ▪ the pitfalls and barriers to effective transition of care from paediatric to adult services <p>Be able to</p> <ul style="list-style-type: none"> ▪ offer managed transitional care in partnership with local adult services, if required ▪ support the young person in transition to adult services ▪ provide nurse specialist continuity of care during the transition phase

Competences: Angio-oedema without weals

Ref	Pathway stage	Competences: Angio-oedema without weals Pathway
21	Initial recognition - NHS direct or self care	<p>Know</p> <ul style="list-style-type: none"> the signs and symptoms of angio-oedema (36) <p>Be able to</p> <ul style="list-style-type: none"> recognise that the child has angio-oedema (36) seek appropriate health professional advice
22	Initial recognition – health professional	<p>Know</p> <ul style="list-style-type: none"> the signs and symptoms of an angio-oedema (36) <p>Be able to</p> <ul style="list-style-type: none"> make an initial assessment recognise that the child has angio-oedema (36) identify any airway compromise and refer for emergency medical help refer onwards for immediate specialist assessment
23	Further specialist assessment and management	<p>Know</p> <ul style="list-style-type: none"> that this care should be provided using a multidisciplinary approach which may involve a paediatric allergist, dermatologist, immunologist, paediatric gastroenterologist, nurse specialist(s), dietitian, dentist, oral surgeon, pharmacist, and psychological support
24	Specialist assessment and management – detailed history/examination	<p>Know</p> <ul style="list-style-type: none"> the causes and exacerbating factors of angio-oedema (34) that angio oedema without weals may indicate a diagnosis of hereditary angio-oedema (HAE) <p>Be able to</p> <ul style="list-style-type: none"> take a clinical history and examination, including gastrointestinal symptoms (34) identify possible precipitating causes (e.g. food, drugs) and aggravating factors (e.g. heat, stress, viral infections) (34)
25	Specialist assessment and management – investigations	<p>Have</p> <ul style="list-style-type: none"> access to appropriate facilities, practical skill and knowledge to undertake and interpret investigations including serum complement, C1 inhibitor and C1 inhibitor function (30) <p>Know</p> <ul style="list-style-type: none"> how to interpret levels of serum complement, C1 inhibitor and C1 inhibitor function (30)
26	Specialist assessment and management – oral-facial granuloma	<p>Be able to</p> <ul style="list-style-type: none"> identify patients with clinical oral-facial granuloma (OFG) presentation treat and refer OFG patients appropriately

Ref	Pathway stage	Competences: Angio-oedema without weals Pathway
27	Specialist assessment and management – treatments	<p>Have access to</p> <ul style="list-style-type: none"> ▪ supplies of C1 inhibitor <p>Know</p> <ul style="list-style-type: none"> ▪ the indications and side effects of prophylactic treatment (e.g. Tranexamic acid, Danazol) (53) ▪ the indications for using C1 inhibitor ▪ the consensus statement for C1 inhibitor deficiency (30) including prophylactic treatment and treatment of acute episodes <p>Be able to</p> <ul style="list-style-type: none"> ▪ initiate prophylactic treatments and monitor patient response ▪ ensure adequate supply of emergency first line treatment (e.g. C1 inhibitor), including selected home availability where appropriate (30) ▪ administer C1 inhibitor in a safe environment ▪ counsel and advise patients and their families on administration of prophylactic and acute treatment(s) ▪ discuss individual circumstances (e.g. administration of C1 inhibitor in light of personal beliefs) ▪ provide an agreed verbal and written personal management plan for treatment and management of further episodes (30)
28	Specialist assessment and management – advice, family screening, minimise quality of life impacts	<p>Know</p> <ul style="list-style-type: none"> ▪ the situations which may exacerbate the episodes of angio-oedema ▪ the indications for providing C1 inhibitor concentrate ▪ the different types of C1 inhibitor deficiency and their patterns of inheritance ▪ the impact on quality of life from a hereditary angio-oedema diagnosis <p>Be able to</p> <ul style="list-style-type: none"> ▪ advise about management of high risk situations including dental surgery ▪ investigate other family members of affected children ▪ liaise with immunology services to provide support for adult family members ▪ provide advice and support for future management, including support the family to reduce the impact on quality of life

Ref	Pathway stage	Competences: Angio-oedema without weals Pathway
29	Specialist assessment and management – Signposting, liaison with schools and early years settings	<p>Know</p> <ul style="list-style-type: none"> ▪ the available support for patients from the <u>Primary Immunodeficiency Association (PiA) (54)</u>, <u>British Association of Dermatologists (42)</u> ▪ the pitfalls and barriers to effective transition of care from paediatric to adult services <p>Be able to</p> <ul style="list-style-type: none"> ▪ provide information for families to access the PiA ▪ liaise with schools and early years settings ▪ liaise with local emergency departments regarding supply of C1 inhibitor concentrate supplies ▪ inform children and families about the process and appropriate timing for obtaining a medical alert talisman (e.g. medical identity bracelet) (53)
30	Specialist assessment and management – transitional care	<p>Have access to:</p> <ul style="list-style-type: none"> ▪ an adult multidisciplinary team <p>Know</p> <ul style="list-style-type: none"> ▪ the pitfalls and barriers to effective transition of care from paediatric to adult services <p>Be able to</p> <ul style="list-style-type: none"> ▪ offer managed transitional care in partnership with local adult services ▪ support the young person in transition to adult services ▪ provide nurse specialist continuity of care during the transition phase

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2. Urticaria Care Pathway: Initial Recognition and Treatment – Self Care.
3. Urticaria Care Pathway: Initial Recognition and Treatment – Health Professional/Nhs Direct.
4. Urticaria Care Pathway: Further Assessment and Management – Detailed History and Examination, Trigger(S) and Aggravation(S).
5. Urticaria Care Pathway: Further Assessment and Management – Investigations.
6. Urticaria Care Pathway: Further Assessment and Management – Adjust Medication/Advice, Monitor Symptoms, Onward Referral.
7. Urticaria Care Pathway: Further Assessment and Management – Quality of Life, Signposting.
8. Urticaria Care Pathway: Specialist Assessment – Multidisciplinary Team
9. Urticaria Care Pathway: Specialist Assessment – History/Examination, Impact on Quality of Life, Trigger(S)/Aggravations.
10. Urticaria Care Pathway: Specialist Assessment – Extended Diagnostic Programme, Test for Physical Urticarias.
11. Urticaria Care Pathway: Specialist Assessment – Adjust Medications/Avoidance Advice, Review and Optimise Treatment.
12. Cutaneous Mastocytosis Care Pathway: Initial Recognition –Self Care.
13. Cutaneous Mastocytosis Care Pathway: Initial Recognition – Health Professional.
14. Cutaneous Mastocytosis Care Pathway: Further Specialist Assessment and Management.
15. Cutaneous Mastocytosis Care Pathway: Further Specialist Assessment and Management – Detailed History and Examination.
16. Cutaneous Mastocytosis Care Pathway: Further Specialist Assessment and Management – Age Appropriate Investigations.
17. Cutaneous Mastocytosis Care Pathway: Further Specialist Assessment and Management – Advice/Information, Management of Further Episodes, Symptoms, Co-Morbidities.
18. Cutaneous Mastocytosis Care Pathway: Further Specialist Assessment and Management – Minimise Impact on Quality of Life, Signposting, Liaison with Schools and Early Years Settings, Long Term Support.
19. Cutaneous Mastocytosis Care Pathway: Further Specialist Assessment and Management – Transitional Care.
20. Angio-Oedema Care Pathway: Initial Recognition – Nhs Direct or Self Care.
21. Angio-Oedema Care Pathway: Initial Recognition – Health Care Professional.
22. Angio-Oedema Care Pathway: Further Specialist Assessment and Management.
23. Angio-Oedema Care Pathway: Further Specialist Assessment and Management – Detailed History/Examination.
24. Angio-Oedema Care Pathway: Further Specialist Assessment and Management – Investigations.
25. Angio-Oedema Care Pathway: Further Specialist Assessment and Management – Oral-Facial Granuloma.
26. Angio-Oedema Care Pathway: Further Specialist Assessment and Management – Treatments.
27. Angio-Oedema Care Pathway: Further Specialist Assessment and Management – Advice, Family Screening, Minimise Quality of Life Impacts.
28. Angio-Oedema Care Pathway: Further Specialist Assessment and Management – Signposting, Liaison with Schools and Early Years Settings.
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