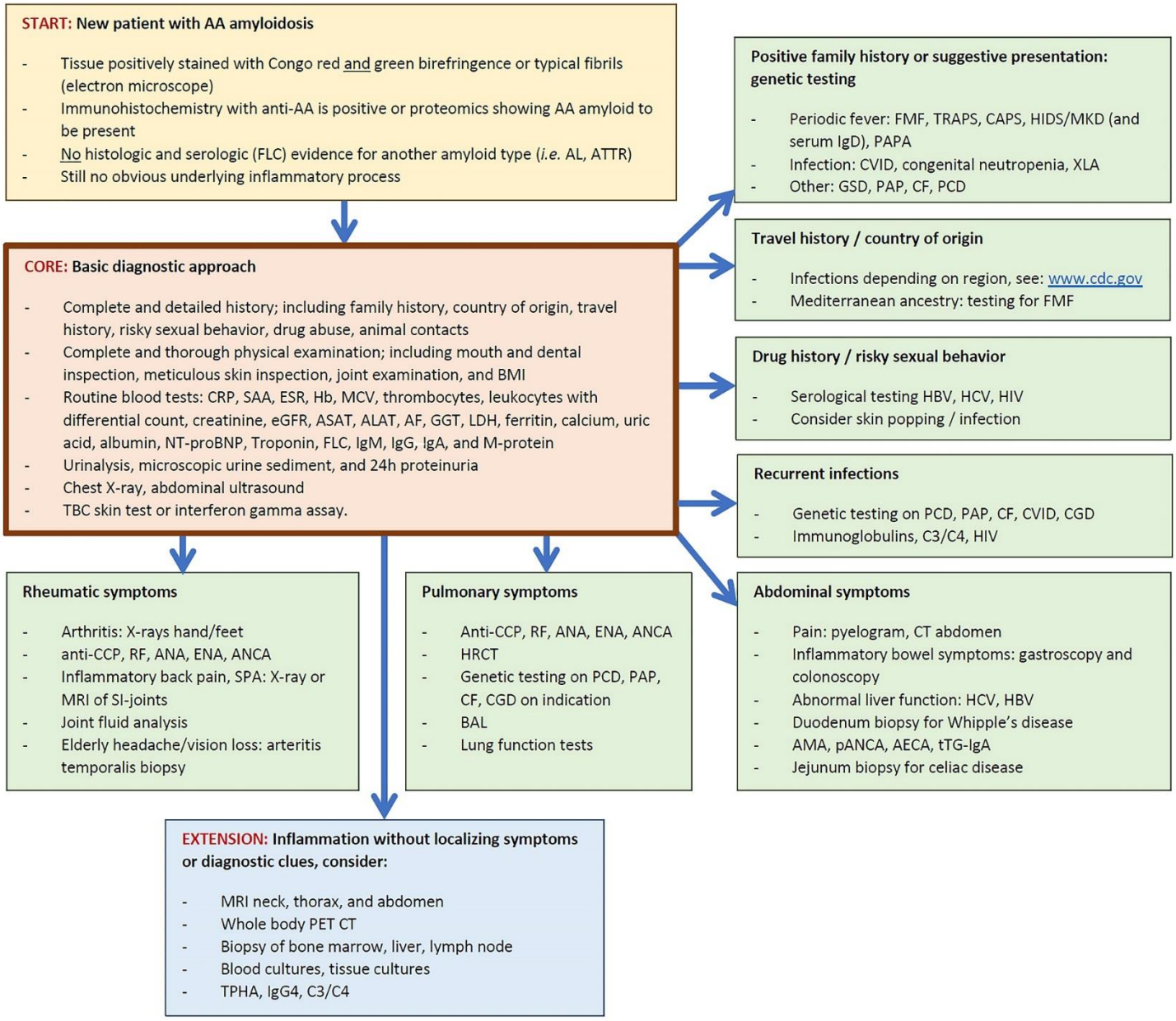
Flowchart detecting causes AA-amyloidosis



Flow chart of the diagnostic approach to detect the underlying disease process in a patient with newly diagnosed AA amyloidosis, but still without an obvious cause (yellow box). First step is obtaining a complete history and performing a thorough physical examination, together with a core set of basic laboratory measurements and imaging (orange box). Second step shows the possible investigations in six directions guided by specific clues obtained from history or from symptoms (green boxes). If localizing symptoms or diagnostic clues remain absent, a full search for the underlying inflammatory process is justified (blue box).

Abbreviations: AA, amyloid A; AECA, anti-endothelial cell antibody; AF, alkaline phosphatase; AL, amyloid light chain-related; ALAT, alanine amino transferase; AMA, anti-mitochondrial antibody; ANA, antinuclear antibody; ANCA, anti-neutrophil cytoplasmic antibody; anti-CCP, anti-cyclic citrullinated protein; ASAT, aspartate amino transferase; ATTR, amyloid transthyretin-related; BAL, bronchoalveolar lavage; BMI, body mass index; C3/C4, complement factor 3 and 4; CAPS, cryopyrin-associated periodic syndrome; CF, cystic fibrosis; CGD, chronic granulomatous disease; CRP, C-reactive protein; CT, computed tomography; CVID, common variable immunodeficiency disorder; eGFR, estimated glomerular filtration rate; ENA, antibodies against extractable nuclear antigen; ESR, erythrocyte sedimentation rate; FLC, immunoglobulin free light chain; FMF, familial Mediterranean fever; GGT, gamma glutamyl transferase; GSD, glycogen

storage disease; Hb, hemoglobin; HBV, hepatitis B virus; HCV, hepatitis C virus; HIDS, hyperimmunoglobulinemia D syndrome; HIV, human immunodeficiency virus; HRCT, high-resolution computed tomography; IgA, immunoglobulin A; IgD, immunoglobulin D; IgG, immunoglobulin G; IgG4, Immunoglobulin G subtype 4; IgM, immunoglobulin M; LDH, lactate dehydrogenase; M-protein, monoclonal protein; MCV, mean corpuscular volume; MKD, mevalonate kinase deficiency; MRI, magnetic resonance imaging; NT-proBNP, N-terminal pro-B-type natriuretic protein; pANCA, perinuclear ANCA; PAP, pulmonary alveolar proteinosis; PAPA, pyogenic sterile arthritis, pyoderma gangrenosum, and acne; PCD, primary ciliary dyskinesia; PET, positron emission tomography; RF, rheumatoid factor; SAA, serum amyloid A protein; TPHA, treponema pallidum haemagglutinase; TRAPS, Tumor necrosis factor receptor-associated periodic syndrome; tTG-IgA, anti-tissue transglutaminase IgA antibodies; XLA, X-linked agammaglobulinemia.