Systemic organ involvement in IgG4-related disease

- Pituitary gland: Headache, visual field deficit, lactation, diabetes insipidus (IgG4-related hypophysitis)
- Lacrimal gland: Swollen upper eyelids, dry eyes (IgG4-related dacryoadenitis)
- Salivary gland: Swollen submandibular portions, dry mouth (IgG4-related sialoadenitis)
- Respiratory tract: Cough; similar to bronchial asthma
- Kidney: Often asymptomatic; hydronephrosis in renal hilum involvement (IgG4-related kidney disease)
- Retroperitoneal cavity: Fever, malaise, aneurysm in cases with periaortitis (IgG4-related retroperitoneal fibrosis)
- Pancreas: Upper abdominal discomfort, obstructive jaundice, impaired glucose tolerance (type 1 autoimmune pancreatitis)
- Lymph nodes: Swollen lymph nodes (IgG4-related lymphadenopathy)

IgG4-related disease can present with various organ dysfunctions. This schematic shows the main organs affected and the clinical features of particular organ manifestations, and outlines the nomenclature used for each organ involvement.
A typical case with IgG4-related disease exemplifying key diagnostic features

a | Dacryoadenitis and sialadenitis manifest as bilateral swelling of the upper eyelids (top panel) and submandibular regions (lower panel), respectively. b | CT images reveal enlargement of the lacrimal gland (top left), submandibular gland (top right), and pancreas (bottom left). Multiple areas of poor contrast enhancement, representative of dense inflammation and fibrosis are seen in the kidney (bottom right). c | A biopsy specimen from the submandibular gland shows severe infiltration of IgG4+ plasmacytes and storiform—whorled or cartwheel patterned—fibrosis (top: haematoxylin and eosin stain, 100× magnification; bottom: anti-IgG4 monoclonal antibody stain with haematoxylin counterstain, 200× magnification).