

IgG4-RD responder index

Date form completed: (e.g., 7 / July / 2050)

Case number: \_\_\_\_\_

Scoring rules

Scoring refers to manifestations of disease activity present in the last 28 days

- Scoring: 0 Normal or resolved  
 1 Improved  
 2 Persistent (unchanged from previous visit; still active)  
 3 New / recurrence  
 4 Worsened despite treatment

Definitions

Organ/site score: the overall level of IgG4-RD activity within a specific organ system

Symptomatic: is the disease manifestation in a particular organ system symptomatic? (Y = yes; N = no)

Urgent disease: disease that requires treatment immediately to prevent serious organ dysfunction (Y = yes; N = no)  
 (presence of **urgent disease** within an organ leads to **doubling** of that organ system score)

Damage: organ dysfunction that has occurred as a result of IgG4-RD and is considered permanent (Y = yes; N = no)

Organ/site	Activity			Damage Present (Yes/No)
	Organ/site score (0-4)	Symptomatic (Yes/No)	Urgent (Yes/No)	
Pachymeninges				
Pituitary gland				
Orbits and lacrimal glands				
Salivary glands				
Thyroid				
Lymph nodes				
Lungs				
Aorta and large blood vessels				
Retroperitoneum, mediastinum, and mesentery				
Pancreas				
Bile duct and liver				
Kidney				
Skin				
Other sclerosis/mass formation				

Descriptor	Level mg/dl	Score
Serum IgG4 concentration		

Total activity score

Organ/sites (×2 if urgent) + serum IgG4 score: \_\_\_\_\_

Steroid dose at the time of assessment (prednisone equivalent):  
 \_\_\_\_ mg/day  
 Cumulative steroid dose in the past 28 days:  
 \_\_\_\_ mg prednisone equivalent

Total number of urgent organs: \_\_\_\_\_

Total number of damaged organs: \_\_\_\_\_

FIGURE 1: IgG4-related Disease Responder Index (IgG4-RD RI) Scoring Sheet this is a sample sheet of the IgG4-RD RI on which physicians score patient's disease activity at a given clinic visit.