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)

	Activity			Damage
Organ/site	Organ/site score (0-4)	Symptomatic (Yes/No)	Urgent (Yes/No)	Present (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		

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

Total activity score

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

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.