

INTERNATIONAL STUDY GROUP OF FABRY NEPHROPATHY SCORE SHEET

Case number:  Scorer:  Date:

**LIGHT MICROSCOPY:** (Only score circled section indicated by a mark.)

1) <b>PODOCYTE VACUOLIZATION</b>		Total	Podocyte score (n affected x score)	Mean Podocyte Score (n affected x score) / total scorable counted
None (score = 0)				
Mild (score = 1; <25%)				
Moderate (score = 2; 25-50%)				
Severe (score = 3; >50%)				

<b>Total scorable glomeruli</b>		<input type="text"/>
Not scoreable, indeterminate (fragment or global GS)		<input type="text"/>
<b>Total number of glomeruli</b> (sum scoreable + indeterminate)		<input type="text"/>

2) <b>OTHER LESIONS</b>		Total
Nonsclerotic glomerulus		
Segmental sclerosis, mild		
Segmental sclerosis, severe		
Global sclerosis		
Adhesion		
Ischemic/collapse		
Peri-glomerular fibrosis and/or Bowman's capsule reduplication		
Indeterminate (fragment of glomerulus)		

3) <b>ARTERIES:</b> Present ___ Absent ___		Total
Average sclerosis (0-3)		
Most severe sclerosis(0-3)		
Hyalinosis:		
	Present ___ Absent ___	
Arterioles:	Medial ___	
	Subendothelial ___	
Arteries:	Medial ___	
	Subendothelial ___	

4) <b>TUBULOINTERSTITIUM</b>		Estimated — scored to nearest 10% (>0 and <5 scored as 5%)
Tubulointerstitial fibrosis		

Interstitial inflammation	
Absent ___	Present ___
	Scarred ___
	Non-scarred ___

**THICK SECTION:** (Only score circled section.)

0; 1 — rare, small; 2 — many, small; 3 — rare, large; 4 — ≥50% large

Number of:	Glomeruli available	Segmental sclerosis		Global sclerosis
		mild	severe	

Inclusions	Glomerulus								Average
	1	2	3	4	5	6	7	8	
Podocyte (0-4+)									

	Present	Absent	Not Sampled
Parietal epithelial inclusions			
Proximal tubular inclusions			
Distal tubular inclusions			
Peritubular capillary inclusions			
Vascular intimal inclusions			
Vascular medial inclusions			

**COMMENTS:**

Revised 05/03/07

Fig. A1. Scoring sheet for Fabry nephropathy by light microscopy.