

A Patient-Focused Study: the Impact of Retroperitoneal Fibrosis

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Introduction

Retroperitoneal fibrosis (RPF) is a rare fibro-inflammatory condition of uncertain aetiology that develops around the abdominal aorta and frequently encapsulates one or both ureter(s) causing pain and obstructive nephropathy. Until recently, gathering information as to how a rare long-term condition such as RPF can impact upon peoples' lives has been difficult.

Objective

To understand the physical and psychosocial impact that RPF has upon peoples' lives

Methods

An international online questionnaire was designed in collaboration with two patient advocates and the multi-disciplinary *Renal Association Rare Disease Registry (RaDaR) RPF Group*. Comprising 62 questions/free text options across 8 sections; demographics, symptoms, medical risk factors, medical sequelae, medical and surgical treatments, psychosocial impact and perceptions of care received, the questionnaire was anonymous and freely accessible on a *GOOGLE Form* online platform for 6 months between 30th September 2016 until 31st March 2017. The survey was advertised through the *RaDaR* website in addition to *Yahoo* and *Facebook* RPF online support groups.

Results summary

Table 1) Demographics. A total of 229 RPF-affected individuals from 30 countries across 5 continents responded.

Mean age at diagnosis (years)	51.4 (range 17-76)
Female (%)	59
Smoker / previous smoker (%)	78
Ethnicity	
White	190 (83%)
Black African / African American	8 (4%)
Hispanic or Latino	8 (4%)
Asian / Pacific Islander	11 (5%)
Other	11 (5%)
Country of origin	
North America	118 (52%)
British Isles	57 (25%)
Mainland Europe	21 (9%)
Australasia	12 (5%)
Asia	9 (4%)
South America	4 (2%)

Figure 1). Worst aspects. Respondents were asked to list their three worst aspects of RPF / its management.

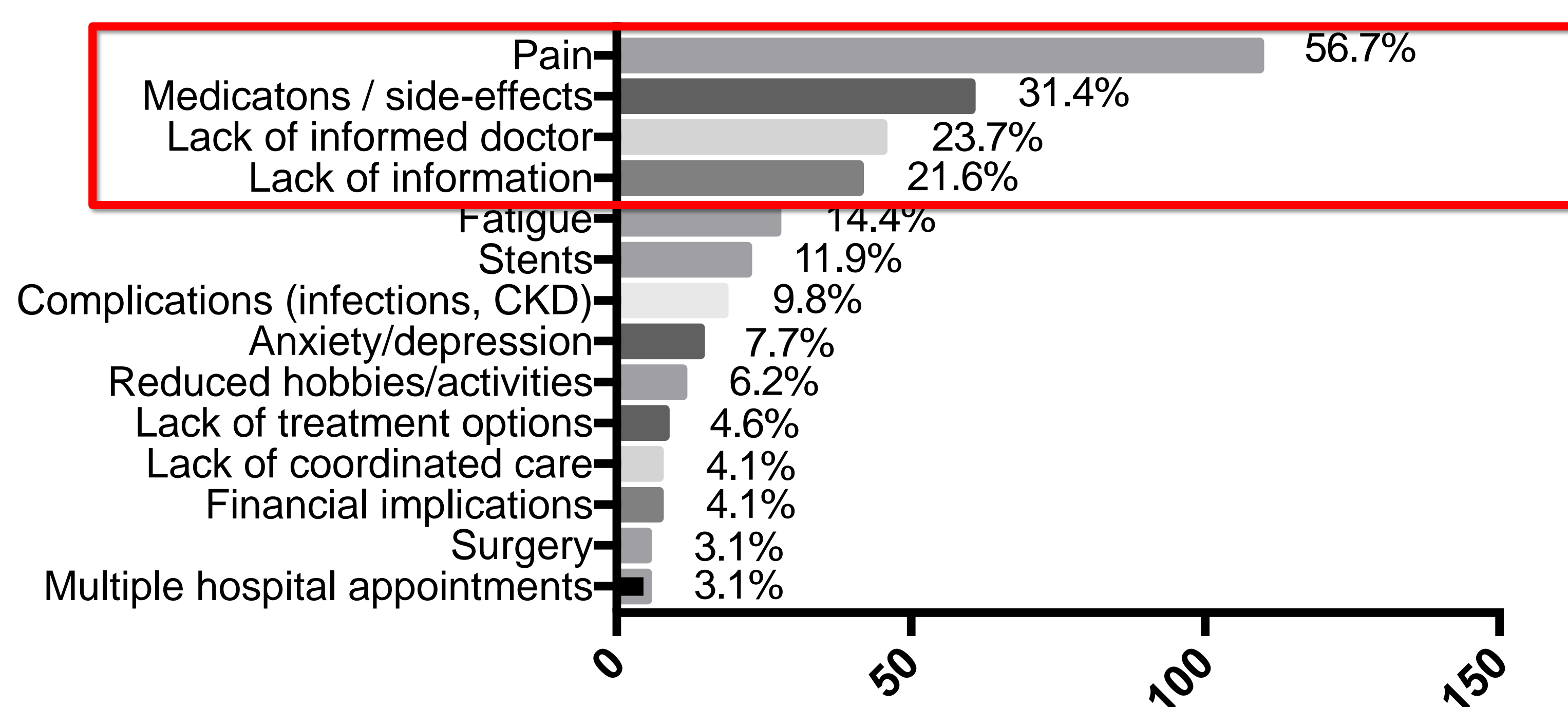


Figure 2) Pain was the 2nd most common physical symptom in RPF with 94.3% reporting taking regular painkillers.

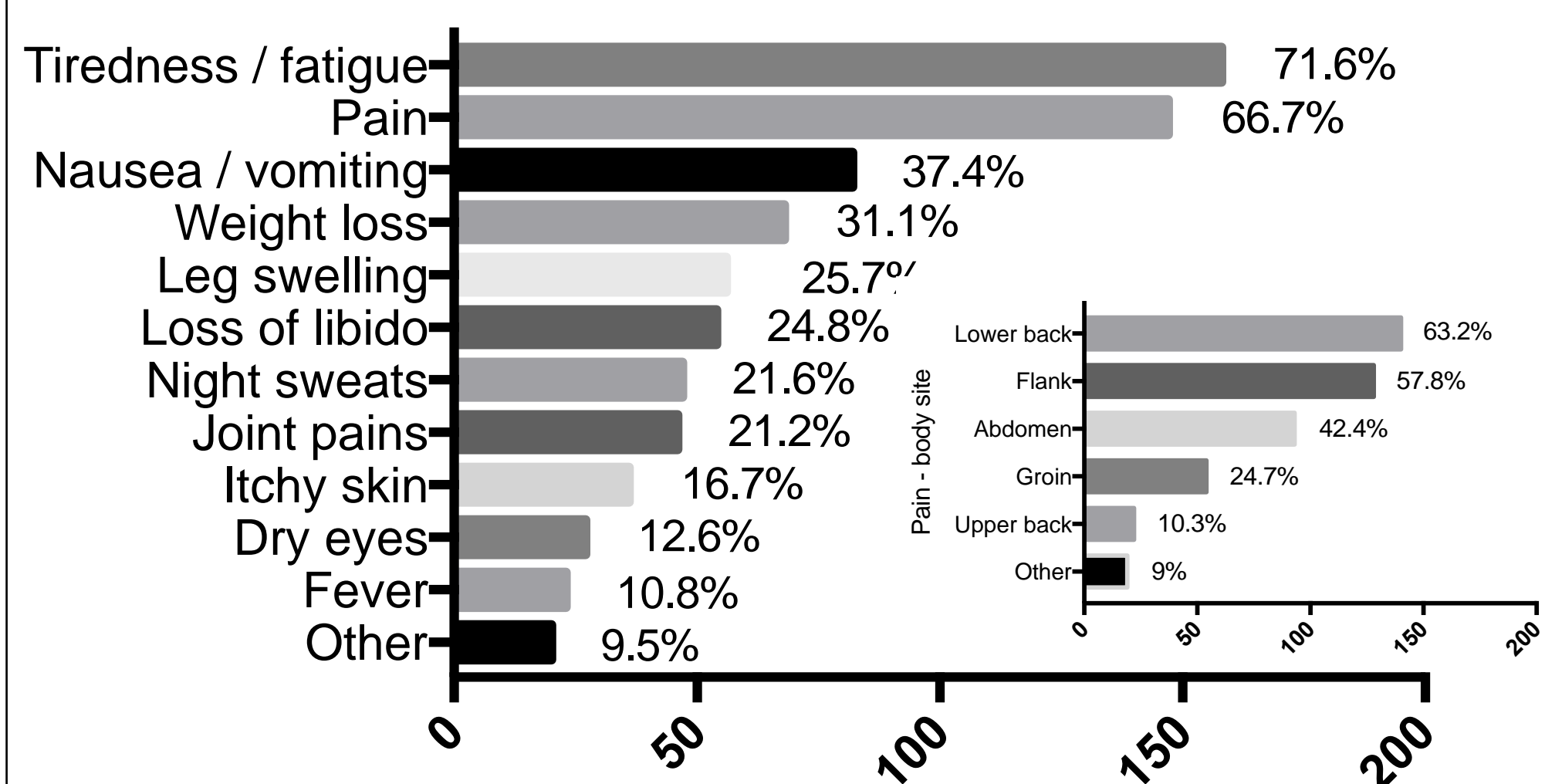
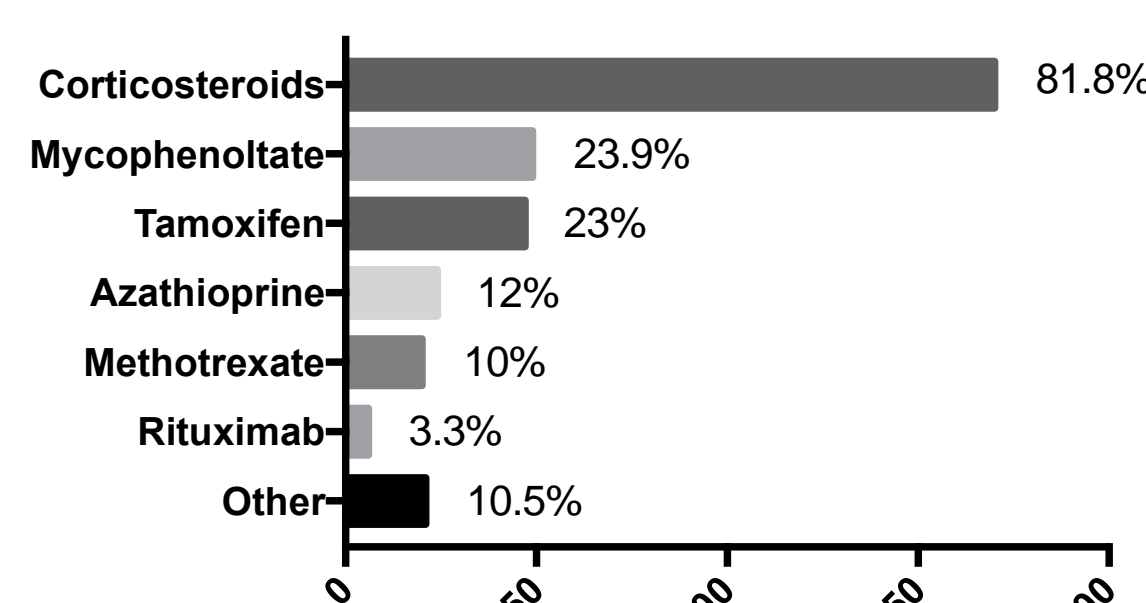
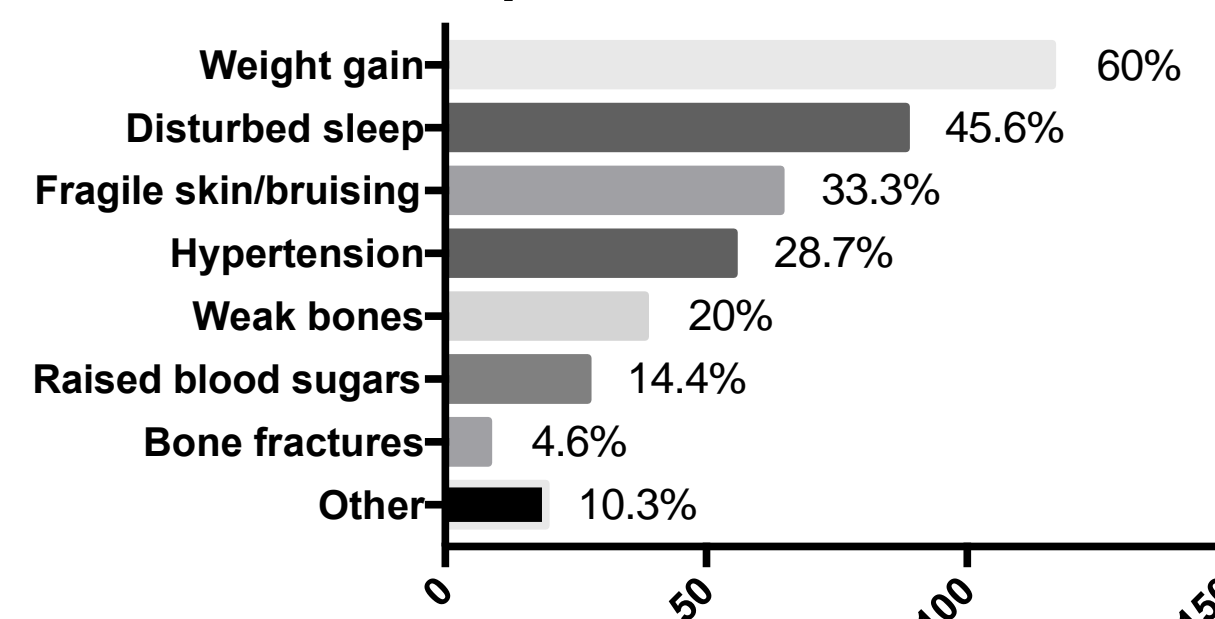


Figure 3) Medications / side-effects.

(a) 81.8% of respondents received corticosteroids as treatment for RPF.



(b) 91.8% of these experienced side-effects



Lack of informed doctor / Lack of information about RPF

Despite 95.2% of respondents seeing more than 2 specialty teams over half:

a) had difficulty in finding a doctor that they felt understood RPF (53.4%)

b) felt that there was no coordination of their care (50.2%)

c) felt their doctor provided enough information about their condition (53.2%)

Respondents ranked their doctors as the third best source of information behind internet searches (33.8%) and internet forums (29.9%).

Conclusion

This study identifies three key issues relevant to the care and well-being of people with RPF; (1) Pain; (2) Medications (including side-effects most commonly relating to corticosteroids); (3) Lack of informed doctors / information about their RPF condition and its management. This information will guide further development of clinical and academic multi-disciplinary activity relating to RPF.

An increasing global usage of online tools and media provides unique opportunities to collect data directly from people suffering from even rare conditions, which will eventually better inform the medical community of unmet healthcare needs and improve patient experience.