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# LETTERS TO THE EDITOR

## Re: 'Diffuse ganglioneuromatosis and plexiform neurofibroma of the urinary bladder: An uncommon cause of severe urological disease in an infant'. J Pediatr Urol. 2013 Apr; 9(3):e131—e133

We read with interest the article by Salvitti et al. [1]. As stated by the authors, visceral involvement is typically insidious, progressive, and difficult to treat. It has been theorized that the plexiform neurofibroma involving the bladder is derived from the vesicoprostatic plexus in males and the urethrovaginal plexus in females [2]. Variable involvement of different segments of the plexus translates into a variable constellation of symptoms and signs that arise as neurofibromas enlarge, compressing and distorting local structures. The unpredictable nature of plexiform neurofibromas has a serious impact on the quality of life of patients with neurofibromatosis type 1(NF1), and their management is challenging for clinicians. Ultimately, the most damaging effect of the tumor is obstruction of the bladder neck or the lower ureter, resulting in unrecognized, progressive renal impairment. This was typified by the case presented by the authors. Most patients with plexiform neurofibroma of urinary tract will eventually develop severe refractory symptoms and require surgery as stated by the authors. However, long-term outcome of surgical management has been disappointing, especially in children with expanding and symptomatic plexiform neurofibromatosis, who must bear the burden of plexiform growth for a lifetime. The contribution of pulmonary arterial hypertension (PAH) that can occasionally complicate plexiform neurofibromatosis and add to high mortality in individual with plexiform neurofibromatosis merits an exploration. Our experience with a 9-year-old girl with plexiform neurofibroma of the genitourinary system (vulva, urinary bladder, and uterus) associated with pulmonary hypertension that was asymptomatic for 9 years but later presented with urinary retention, hypertension, and cardiac failure illustrates this fact [3].

PAH in patients with NF1 is hypothesized to be secondary to an underlying vasculopathy [4]. NF1-associated vasculopathies are heterogeneous and appear to contribute to the mortality of children and young adults [4].

The prognosis of patients with von Recklinghausen disease can be generally good except when complicated by pulmonary hypertension [4]. All four patients with PAH reported by Stewart et al. [4] died of either respiratory failure or presumed cardiac failure. The 9-year-old girl we reported also died of cardiac failure [3]. Pulmonary hypertension is therefore a major determinant of the morbidity and mortality in patients with von Recklinghausen disease. The patient presented by the authors will therefore benefit from further evaluation and monitoring to exclude pulmonary hypertension or detect it early for an improved long-term outcome.

## References

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## Response to Nasir et al.

We sincerely appreciate the comments made by Nasir et al. regarding our paper. The aim of our article was to focus the attention on the involvement of the urinary bladder in patient with NF1.

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