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News Letter

Resolving the Puzzle of Loin Pain and Haematuria Syndrome (LPHS) by Discovering its Overlooked Link with Symptomatic Nephroptosis (SN)

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NEWS LETTER

The loin pain haematuria syndrome (LPHS) was first reported in 1967 [1]. Symptomatic nephroptosis (SN) has been known for centuries [2] but was disparaged over 70 years ago [3]. The link between the two conditions is a new discovery [4,5]. This discovery was based on a prospective observational study of 190 patients of whom 36 suffered from LPHS [5]. It was demonstrated that renal pedicle stretch causes neuro-ischaemia as evidenced by the new IVU 7 sign (Figure 1). Renal sympathetic denervation and nephropexy has proved curable for both LPHS and SN.

All patients presenting with loin pain with or without hematuria during 10 years were entered into a prospective observational study and underwent thorough clinical, laboratory and imaging investigations. Repeated standard imaging including CT, MRI, MRA was invariably normal, when supine. However, 190 patients demonstrated SN of > 1.5 vertebrae (>5 cm) on repeating intravenous urography (IVU) with erect film. Of whom 36 (18.9%) patients developed recurrent episodes of painful hematuria for which no organic pathology was detected on all imaging, when supine- thus fitting the definition of LPHS. The IVU 7 sign, with its horizontal and vertical segments, represents the renal pedicle at supine and erect IVU films, respectively was used for measuring renal pedicle stretch causing renal vessels stenosis and ischemia (Figure 1).

Of 190 with SN on IVU-E, 182 were females and 8 males. The mean age was 28.8, duration of symptoms 15.7 and hospital follow up 6.6 years. All patients showed no abnormality on IVU or ancillary imaging when supine. All patients showed renal drop of >1.5 vertebrae (>5 cm) on erect IVU film. Other demonstrable pathology on erect IVP film included: pelviuretric junction kink affecting the right kidney in 116 (61.1%) and bilateral in 19 (10%) of patients. Stretch/rotation of renal pedicle causing neuro-ischaemic pain of LPHS was demonstrable on the right side in 72 (37.9%) and bilaterally in 7 patients.

Complications of SN on IVU erect film included both obstructive and neuro-ischaemic: obstructive complication included ballooned renal pelvis, hydronephrosis and upper pole diverticulum. Neuro-schaemic complications induced by pedicle stretch and rotation/twist were haematuria of the LPHS affecting 36 (18.9%), auto nephropexy affecting 12 right kidneys, upper pole calyctiasis with extra-vasation affecting 28 (15.8%) right kidney and 2 bilateral that are best shown on RGP. Renal atrophy affected 4 right kidneys. Upper pole infarction affected 2 kidneys. Retrograde pyelography (RGP) also demonstrated upper pole calyctiasis with extra-vasation- the same site of haematuria. Surgical treatment was used in 28 patients; 10 had simple nephropexy and 18 had renal sympathetic denervation and nephropexy (RSD&N) for severe LPHS. Four of patients treated with simple nephropexy had recurrence of LPHS while those who had RSD&N were all cured of both LPHS and SN.

The reported data demonstrate that LPHS is caused by SN. The stretch of renal pedicle on erect posture causes severe stenosis of renal vessels as confirmed by exploration of the

Figure 1 Shows renal pedicle mapped on a supine IVU film (Horizontal) and erect film (Vertical) limbs of 7 where the renal pedicle is stretched to 3 times its normal length, causing stenosis and ischemia.

kidney vessels (Figure 2). Early in LPHS renal vessels stenosis occur intermittently on stretch of erect posture only. Later with chronicity sessels stenosis became organic and fixed. The IVU 7 sign is a unique discovery that demonstrates the underlying pathology of SN causing LPHS via severe renal vessel stenosis and ischemia. This effect is also demonstrated by the tube stretch hypothesis (Figure 3) which shows that stretching a tube to twice its length obliterates its lumen and prevents any flow inside the lumen.

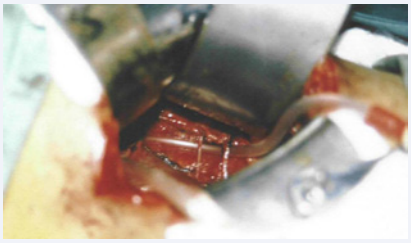

Figure 2 Shows an explored right renal pedicle after denervation of LPHS patient with severely stenosed artery and vein (effect of chronic repeated pedicle stretch). The Nelaton tube is approximately the size of a normal renal artery.

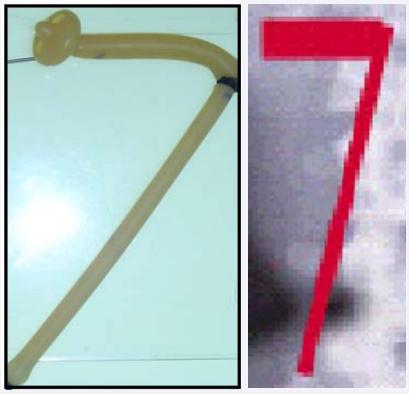

Figure 3 Compares the rubber tube stretch with IVU 7 sign which demonstrate that stretch of a tube or artery causes stenosis of the lumen diminishing flow inside lumen.

Renal sympathetic denervation and nephropexy is a unique surgery has proved most effective and curative for both SN and LPHS. It has transformed the life of these young patients from suffering pain and misery to a quality comfortable and happy life.

CONCLUSION

Upright IVU film and RGP are essential for the diagnosis of SN complicating into LPHS. The new IVU 7 sign affirms that pedicle stretch causes ischaemic nephropathy. Renal sympathetic denervation and nephropexy is curable for LPHS but simple nephropexy is not.

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