

## Medical history

The patient's medical history started 3 months after an accident when the patient fell onto her left thorax while doing homework. After some weeks a very severe left-sided epigastric and thoracic pain radiating towards her left breast developed. This pain persisted and increased and is interfering substantially with the quality of life.

Moreover, a second area of pain is located left to the umbilicus and slightly cranial to it.

During the workup of the abdominal complaints a colonoscopy was carried out which detected early, localized only, anal cancer. The polyp was removed, and the patient was treated with radiation and chemotherapy. So far, no metastases and no local recurrence of the tumor have been demonstrated during the regular checkups.

The patient denies headaches, swelling of the legs, pain in the lower abdomen and loss of consciousness. The symptoms are less while lying supine but increase substantially after standing for a while but even more while driving a car in the sitting position.

The patient started a vegetarian or vegan diet and reduced her weight substantially. This may have been associated with increasing pain.

The further workup showed some hints to a coeliac artery compression and a nutcracker syndrome.

After the consultation of numerous specialists, a causal relationship to her symptoms was denied.

Since some of the complaints have been related by the patient and her treating physicians to breast implants, they have been removed but unfortunately without any effect on the symptoms below the left breast.

## Clinical findings

The patient has unremarkable scars after removal of her breast implants. Below her left breast and along the circumference of the left thorax no cutaneous abnormalities, no lump and no vascular bruit can be heard or felt.

The abdominal palpation reveals pain in the epigastric area and left to the umbilicus but no pathologic lump. A vascular bruit is not audible. The cutaneous vasculature is unremarkable. There are no definite signs of a hyperdistensible connective-tissue. Only the wrist sign is positive, and the elbows can be overstretched.

## Sonographic findings

The urinary bladder is very well filled and unremarkable. Its wall is smooth. The uterus can be easily depicted and shows no congestion. There is no free fluid in the lower abdomen.

After micturition the urinary bladder is empty, and the uterus is still without any signs of congestion.

The left common iliac vein is found substantially compressed while crossing the spine with a maximum diameter of the left to the sacral bone of 12 mm and a minimum diameter at the crossing of the left common iliac vein with the right common iliac artery varying between 1 and 4 mm in a sagittal view.

The flow velocity within the left common iliac vein increases due to its compression from 9 to 41 cm/s. During inspiration an increased perfusion can be demonstrated and the narrowing of the compressed vein enlarges slightly.

Similarly, the right common iliac vein is asymmetrically compressed by the right common iliac artery. The effect is less than on the left side. The flow acceleration is found increasing from 20 to only 30 cm/s.

In a supine position a strong lordosis can be demonstrated. The minimum distance between the ventral surface of the fourth lumbar vertebra and the inner lining of the abdominal wall is decreased to 18 mm

only.

This lordosis produces a slight compression of the vena cava inferior with a flow acceleration in a supine position from 54 to 130 cm/s. The fastest flow can be found slightly cranial to the apex of the lumbar curvature and is produced by the compression by a bowel loop in front of the uplifted spine which slightly compresses the vena cava to produce the above-mentioned flow acceleration.

The most severe effect of the lordotic curvature is found with the left renal vein. This vessel is completely compressed between the superior mesenteric artery and the aorta. No blood flow can be demonstrated crossing this clamp and the entire blood is diverted via a trunc réno-rachidien towards the hemiazygos vein. This vessel is found to be enlarged up to the level of the 10<sup>th</sup> lumbar vertebra. Here the patient feels some tension and discomfort while demonstrating the course of the vein along her spine. The sensation is strictly different from the pain which radiates from the epigastric angle towards the caudal circumference of her left breast.

Moreover, the origin of the coeliac trunk is stretched and distorted by a tightly straddling claw-like arcuate ligament. Within the aorta the flow velocity is 61 cm/s and the flow volume above the aortic hiatus in a supine posture is 2797 mL/min.

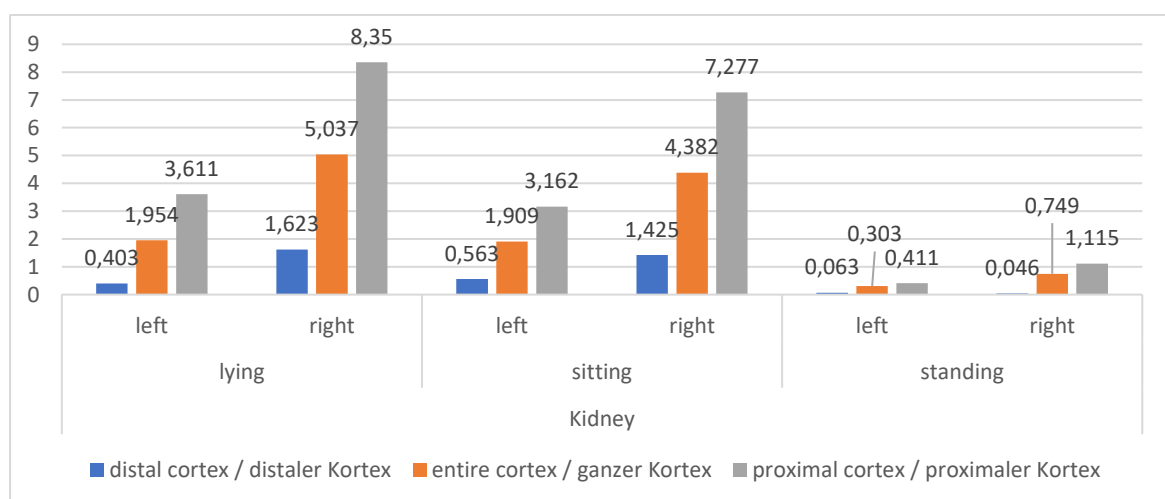
In the midposition of the diaphragm the flow acceleration underneath the tightly straddling median arcuate ligament is found at 315 cm/s and drops to 213 cm/s in deep inspiration. In expiration the flow velocity increases again to 304 cm/s. This constitutes a clinically relevant median arcuate ligament syndrome with the typical clinical findings of pain which radiates from the compression site towards the left lower thorax. This conclusion can be substantiated by the improvement of the pain below the left breast while pressing the thorax from both sides which typically reduces the impact of the median arcuate ligament onto the coeliac plexus. Thus, in accordance with the clinical findings and the patient's symptoms the ultrasound provides a clear diagnosis of a coeliac artery compression syndrome.

Since the symptoms are dependent on the body posture the position of the kidneys in supine and sitting and standing positions are compared. Both kidneys sink caudally while the trunk is upright but the distance on both sides is only 1.3 cm. Nevertheless, the blood flow changes dramatically while standing but does not drop while sitting. In all positions the left kidney has a strong blood flow reduction to 1/3 to 1/2 of the right side-this can be shown in the diagram below. The constant deterioration of the pain while driving a car can thus not be related to the nutcracker syndrome of the left renal vein despite the incomplete compensatory effect of the bypassing of venous blood towards the hemiazygos vein. The left ovarian vein is not found to be a collateral pathway of the left renal vein.

Thus, the pain during driving a car is attributed to the median arcuate ligament syndrome.

### Diagram of the renal parenchymal perfusion measurement with the PixelFlux technique

The columns indicate the perfusion intensity in cm/s calculated as perfusion velocity [cm/s] of all colored pixels multiplied by the area of all colored pixels [cm<sup>2</sup>] divided by the area of all pixels of the entire region of interest [cm<sup>2</sup>]



The duodenum is not compressed while crossing the aorta. The mesenteric artery lies right to the aorta and not in front of the largest artery. Thus, a compression of the duodenum cannot be exerted by both

arteries. A Wilkie syndrome can be ruled out.

Both kidneys have a normal appearance. The left kidney is enlarged to 89 mL/m<sup>2</sup> as a consequence of its venous congestion whereas the right kidney has a normal volume of 71 mL/m<sup>2</sup>.

The blood flow volume running through the tronc réno-rachidien is about 200 mL/min which is not sufficient to provide a fully compensatory collateralization. On the other hand, only the pain in the left mid-abdomen can be attributed to the congestion of the left renal vein.

The blood flow within the distal external iliac vein on both sides changes only a little while standing. On the left side during a supine position the flow volume is 277, on the right side 158 mL/min. While standing at the same location the flow drops on the right side to 38 mL/min whereas on the left side the flow is only little reduced to 210 mL/min.

The left thorax is scanned to demonstrate any pathology of the thoracic wall. No abnormality can be demonstrated.

Moreover, the echocardiography does not reveal a reduced left ventricular function-ejection fraction is 62%.

The aortic flow volume only changes slightly while standing to 2683 mL/min while the heart frequency increases to 105 bpm. No clinically relevant pooling of venous blood within the pelvis and no postural tachycardia can be diagnosed from these findings.

## Diagnoses

1. Severe and clinically dominant median arcuate ligament syndrome which is responsible for the epigastric pain radiating towards the left breast
2. Complete compression of the left renal vein due to the lordogenetic uplifting of the aorta
3. Insufficient collateralization of the compressed left renal vein via a tronc réno-rachidien towards the left hemiazygos vein
4. No pelvic congestion
5. Low-grade and clinically irrelevant May- Thurner constellation
6. Even lesser asymmetric compression of the right external iliac vein producing a pressure gradient with a pressure increase by the factor of 1.5
7. Low-grade lordogenetic compression of the vena cava while lying supine which disappears in positions with uplifted trunk
8. Severe lumbar lordosis
9. No Wilkie syndrome
10. No left ventricular dysfunction, no dilatation, no pericardial effusion, no coarctation of the aorta
11. No relevant connective-tissue disorder with respect to the symptoms of the patient

## Recommendations

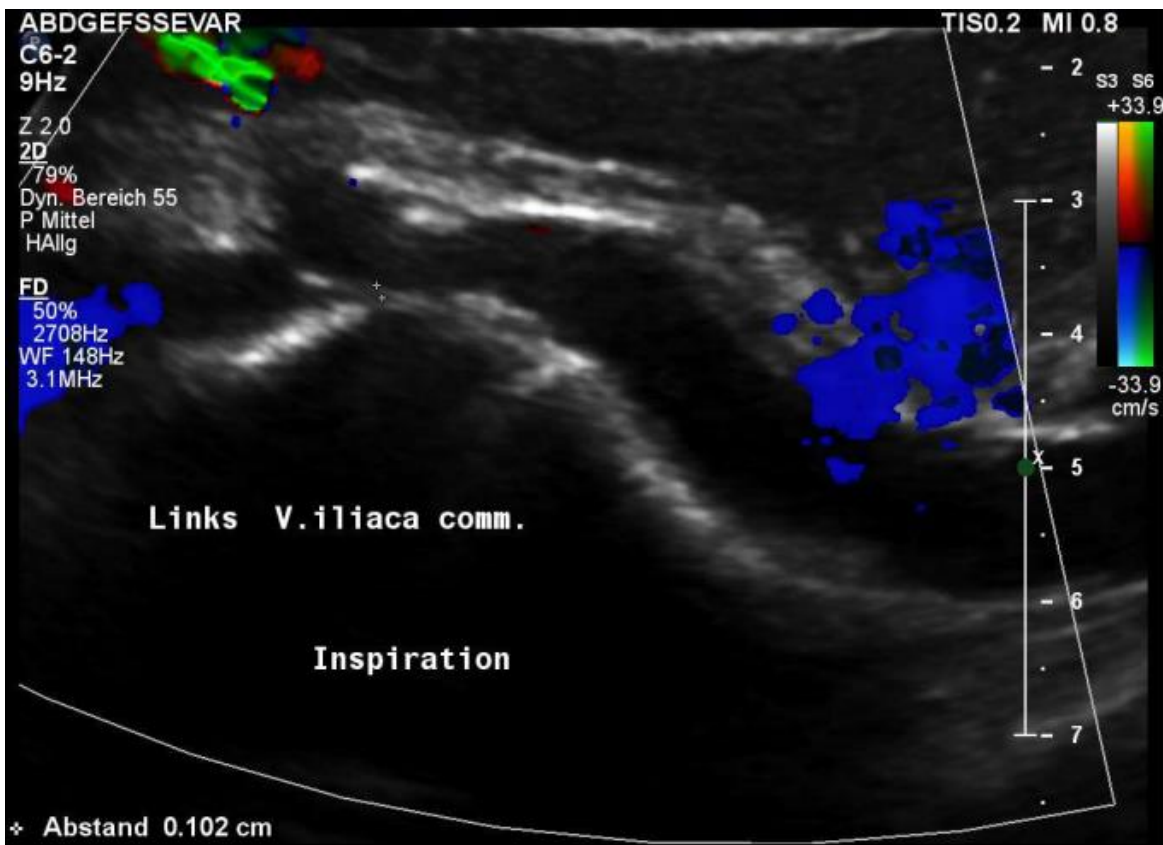
I recommend starting physiotherapy, yoga with breathing exercises and muscle training to increase the strength of the abdominal muscles and deep muscles of the back in order to reduce to lordosis of the lumbar spine which is the main cause of both vascular compressions. An attempt should be made to reduce the venous congestion which is responsible for the pain in the left mid abdomen by administration of 100 mg aspirin for 3 months.

I would expect a reduction of the pain in the left flank but not a fundamental effect onto the pain in the lower left thorax and in the epigastric angle.

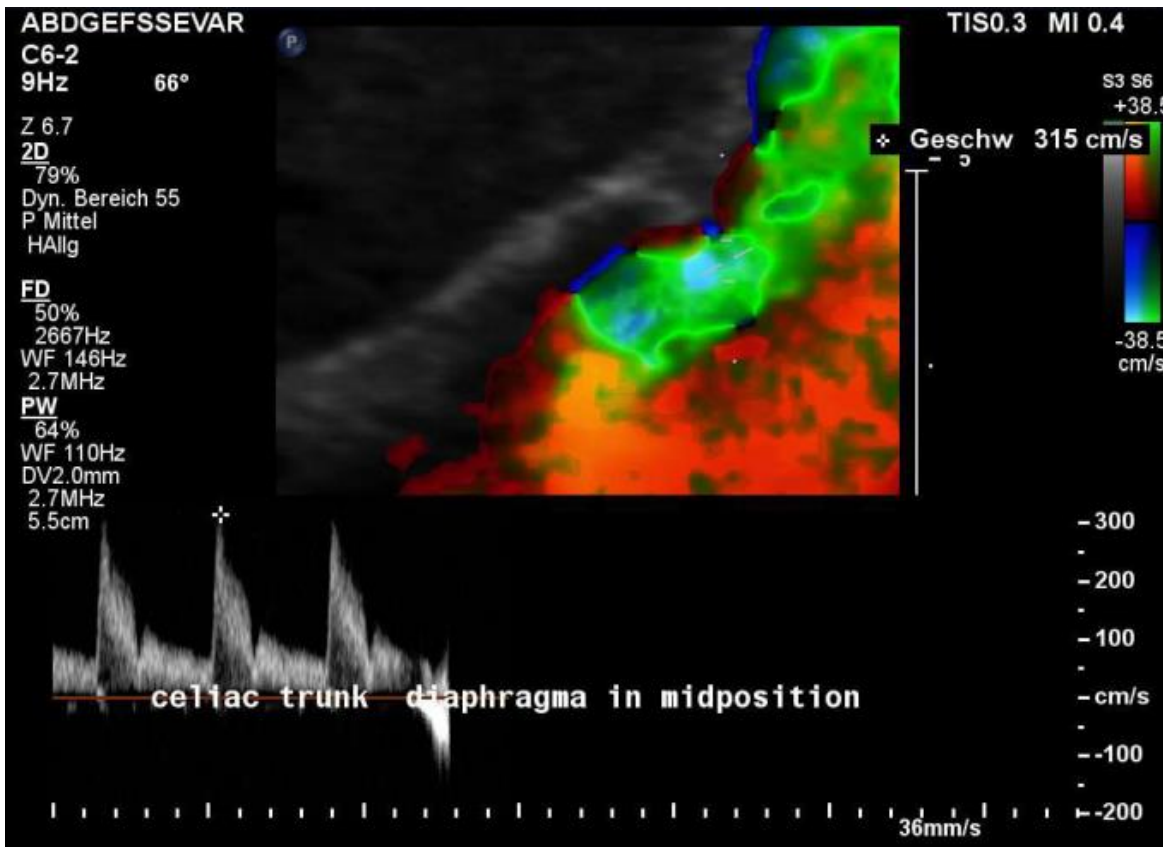
Here I recommend a surgical decompression of the coeliac plexus.

Professor Dr. med. habil. Thomas Scholbach

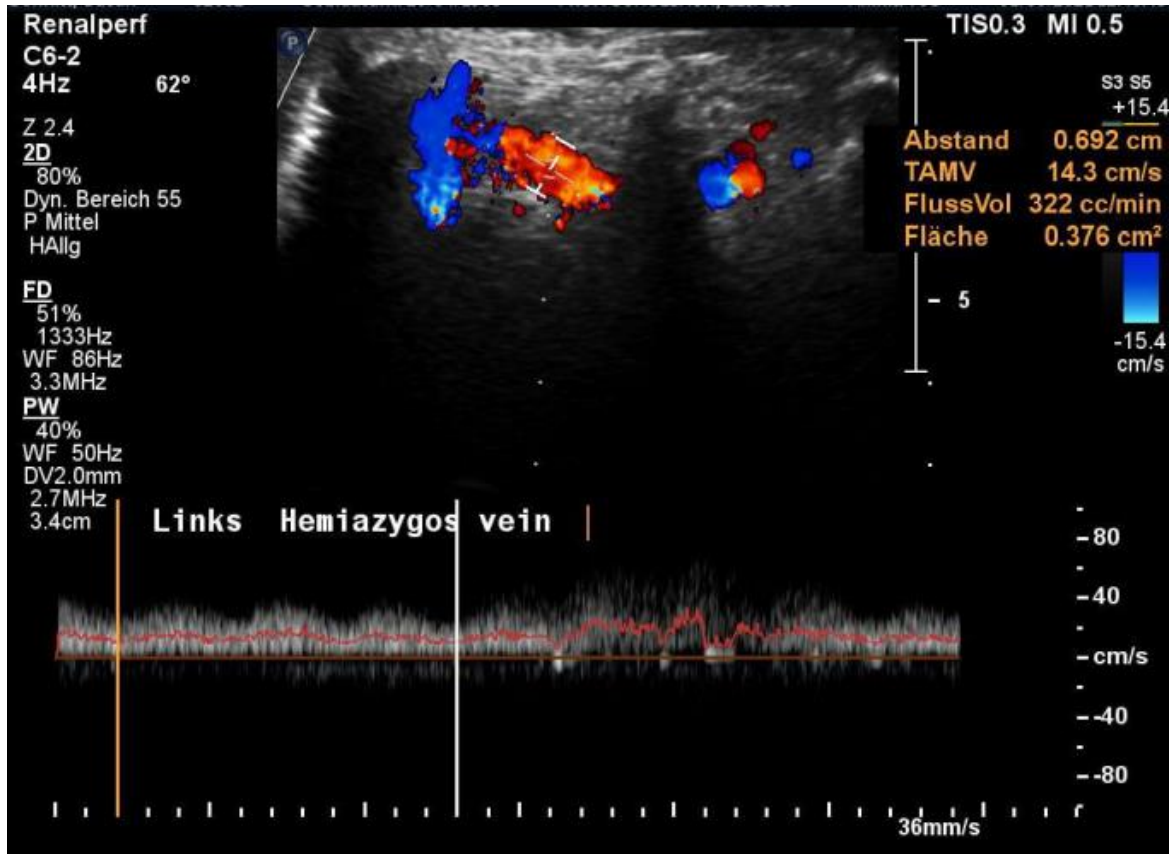
Illustrative images



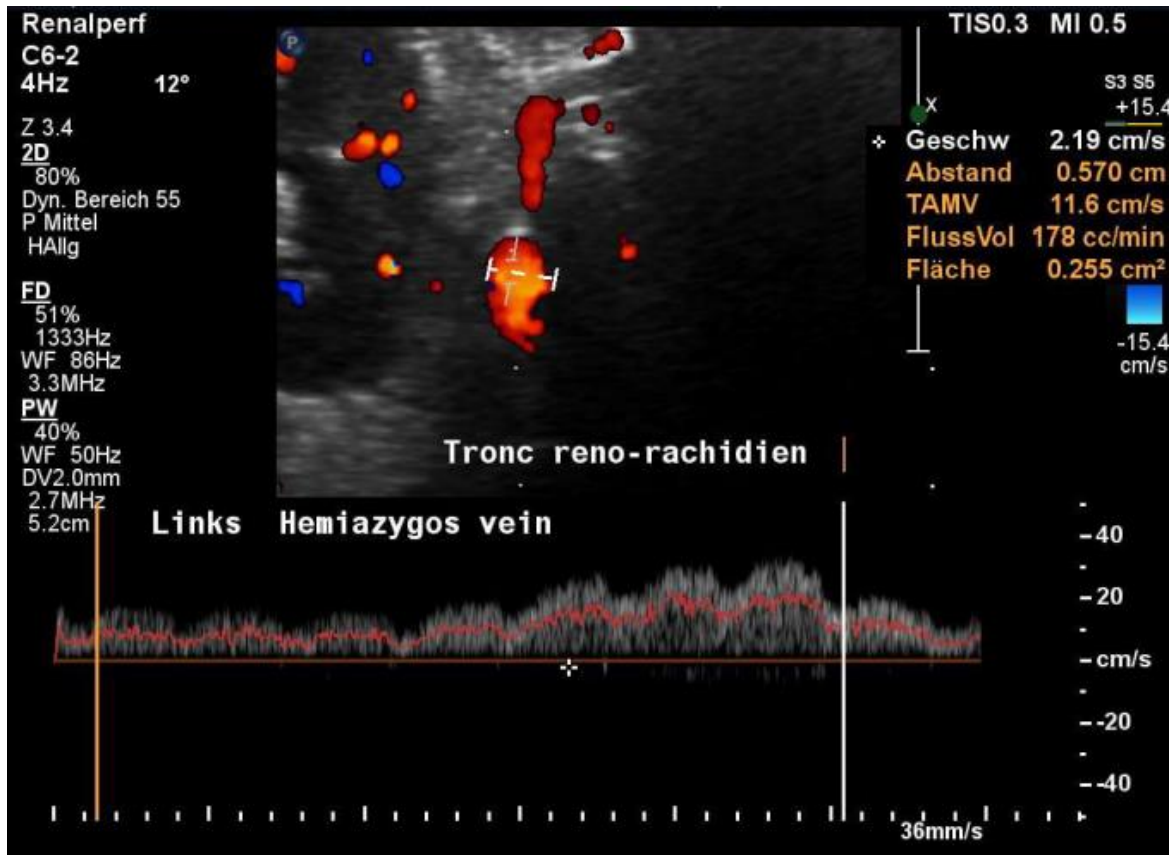
May-Thurner-constellation even in inspiration



Coeliac trunk compression



Increased perfusion of the hemiazygos vein as part of the collateralization



Tronc reno-rachidien connecting the left renal vein with the veins of the spine