IMAGES in PAEDIATRIC CARDIOLOGY

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Abstract
Delineation and documentation of anatomy in the presence of significant mass pathology presents a diagnostic challenge. This often necessitates the implementation of more than one imaging modality in order to perform an adequate assessment. We present a three-year old boy with extensive distortion of mediastinal anatomy secondary to pleural metastases from a Wilms tumour. This limited the ability to accurately assess mediastinal anatomy and cardiac function at baseline. Reassessment following initiation of chemotherapy showed a significant reduction in size of metastases with complete resolution of the mediastinal distortion.

MeSH
Wilm's tumour.

Case presentation
We present a three-year old boy who presented to the Emergency Department at Mater Dei Hospital in Malta with respiratory distress and abdominal distension on a background of a one-month history of cough. On assessment, he was tachypnoeic with decreased air entry and dullness to percussion on the right side of the chest. Abdominal examination revealed a left sided flank mass. A chest radiograph showed opacification of the entire right lung field with mediastinal shift to the left and a possible retrocardiac mass (figures 1 and 2).

Figure 1: CXR showing opacification of the right lung field

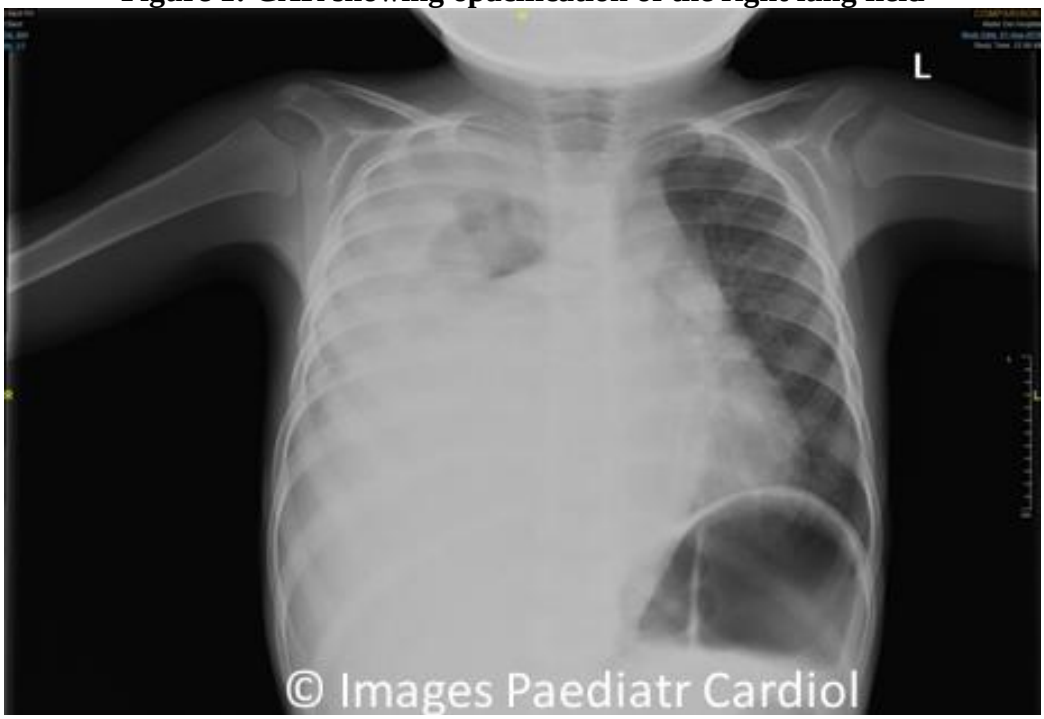
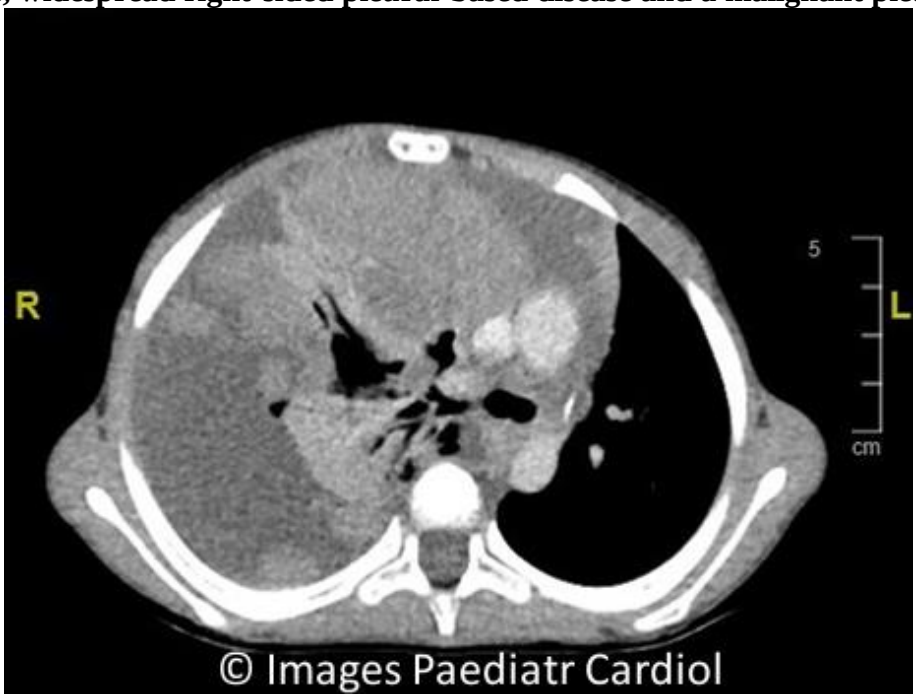


Figure 2: Computed tomography (CT) imaging of the trunk (bottom) showing a left renal tumour mass, widespread right-sided pleural-based disease and a malignant pleural effusion.



Echocardiography revealed a heart that was significantly displaced (posteriorly and leftwards) by the intrathoracic mass. intracardiac structures and connections were normal. There was resultant distortion of the course of the great vessels, with the abdominal aorta and inferior vena cava being displaced anteriorly, navigating their way around the mass. This circumvented course hindered full interrogation of the vessels, and it was difficult to assess patency or presence of tumour invasion in the proximal segments of these vessels. The right atrium was externally compressed by the intrathoracic mass, rendering the right atrium to a slit-like shape, without any obvious obstruction to venous inflow. The significant cardiac displacement did not allow correct measurements of cardiac function such that indices relying on correct angulation could not be used (Fig 3-5).

Figure 3:Echocardiogram showing tumour compressing right atrium (red arrow)

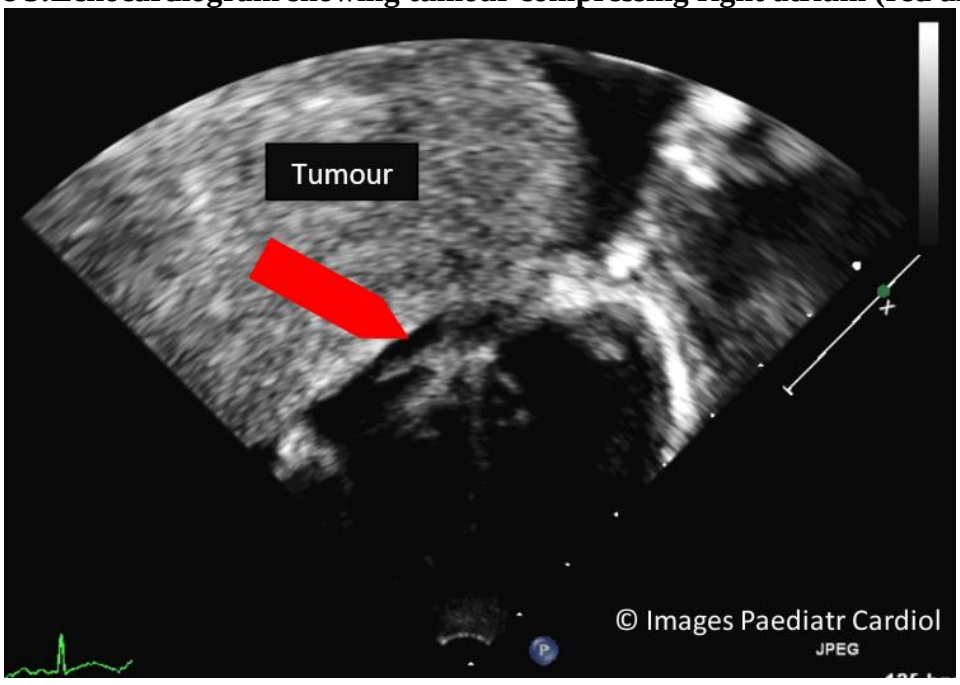


Figure 4: Echocardiogram showing tumour compressing left atrium (red arrow)

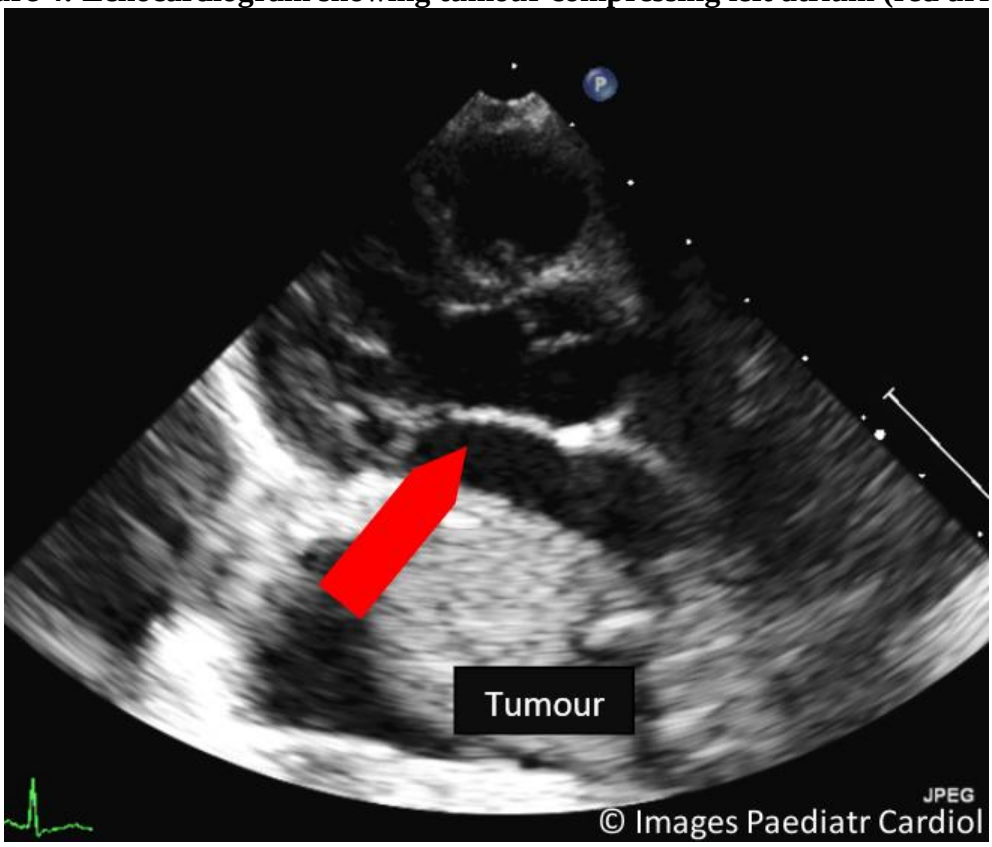
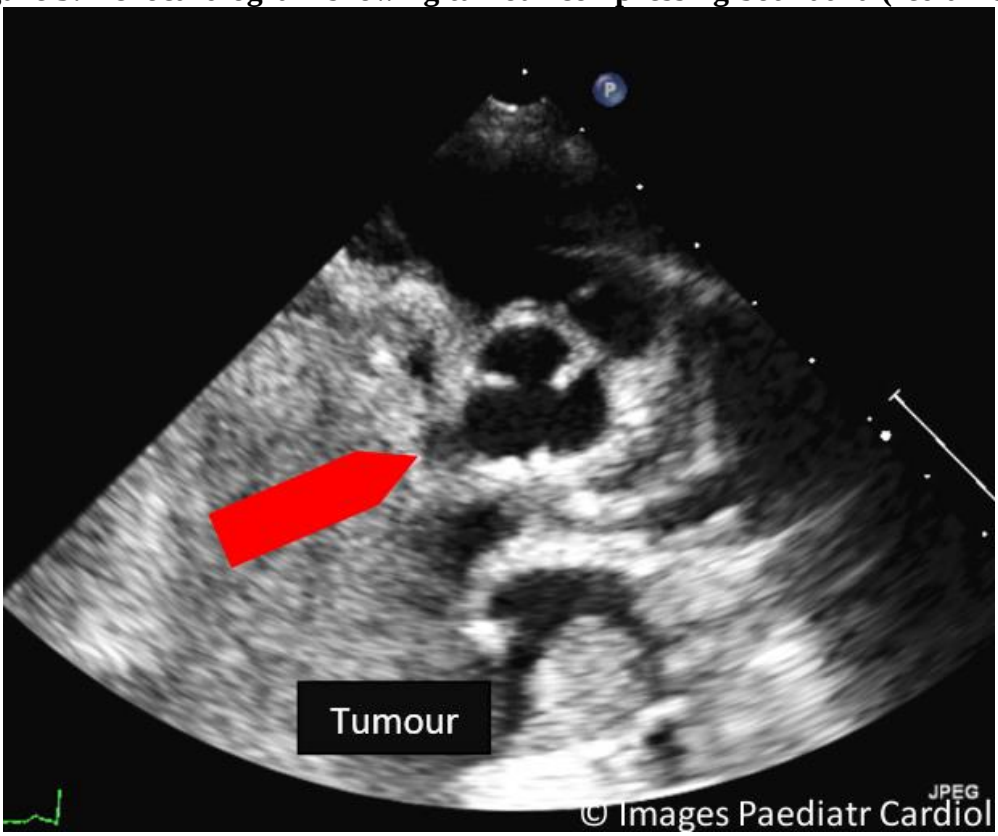


Figure 5: Echocardiogram showing tumour compressing both atria (red arrow)



After histological confirmation of nephroblastoma of the left kidney, chemotherapy with intravenous Vincristine, Actinomycin D and Doxorubicin was commenced according to the Clinical Management Guidelines Wilms Tumour by the Children's Cancer and Leukaemia Cancer Group.

The clinical condition improved and the respiratory distress gradually resolved. Magnetic resonance imaging (MRI) with venous studies performed just two weeks after admission showed a significant reduction in the size of the renal primary and the right pleural based and mediastinal mass with resolution of the mediastinal shift, with no inferior vena cava or left renal vein thrombus. Chest radiography performed three weeks after admission showed improved aeration of the right lung and reduction in the size of the pleural effusion. Repeat echocardiography four weeks after admission showed improved findings with resolution of the previously documented cardiac distortion. Normal function could now be documented clearly in view of correct angles of measurement and normal acoustic windows. (Figs 6-8).

Figure 6: Chest radiograph showing significant reduction in right-sided pleural-based disease.

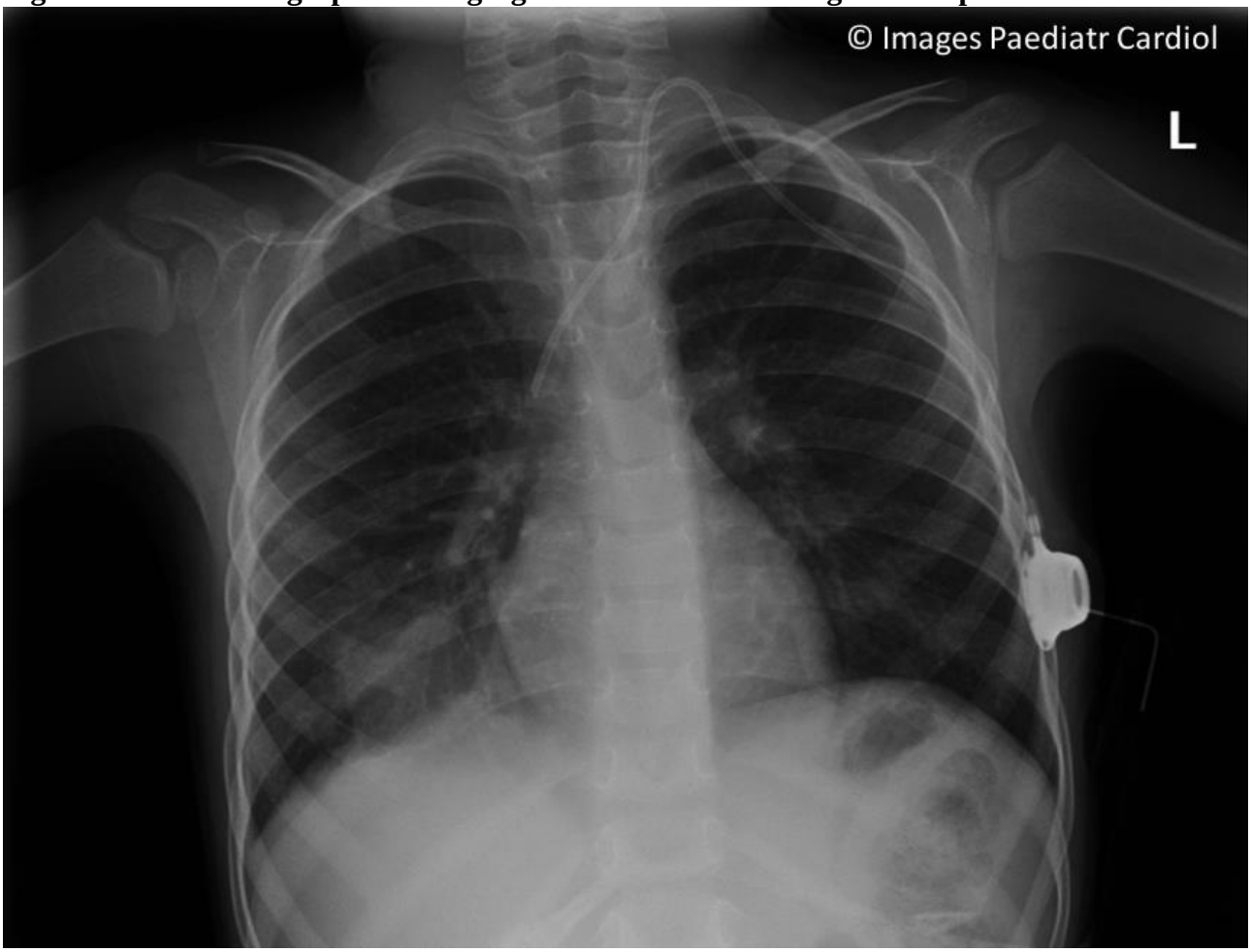


Figure 7: Echocardiography showing relief of pressure and re-expansion of atria.

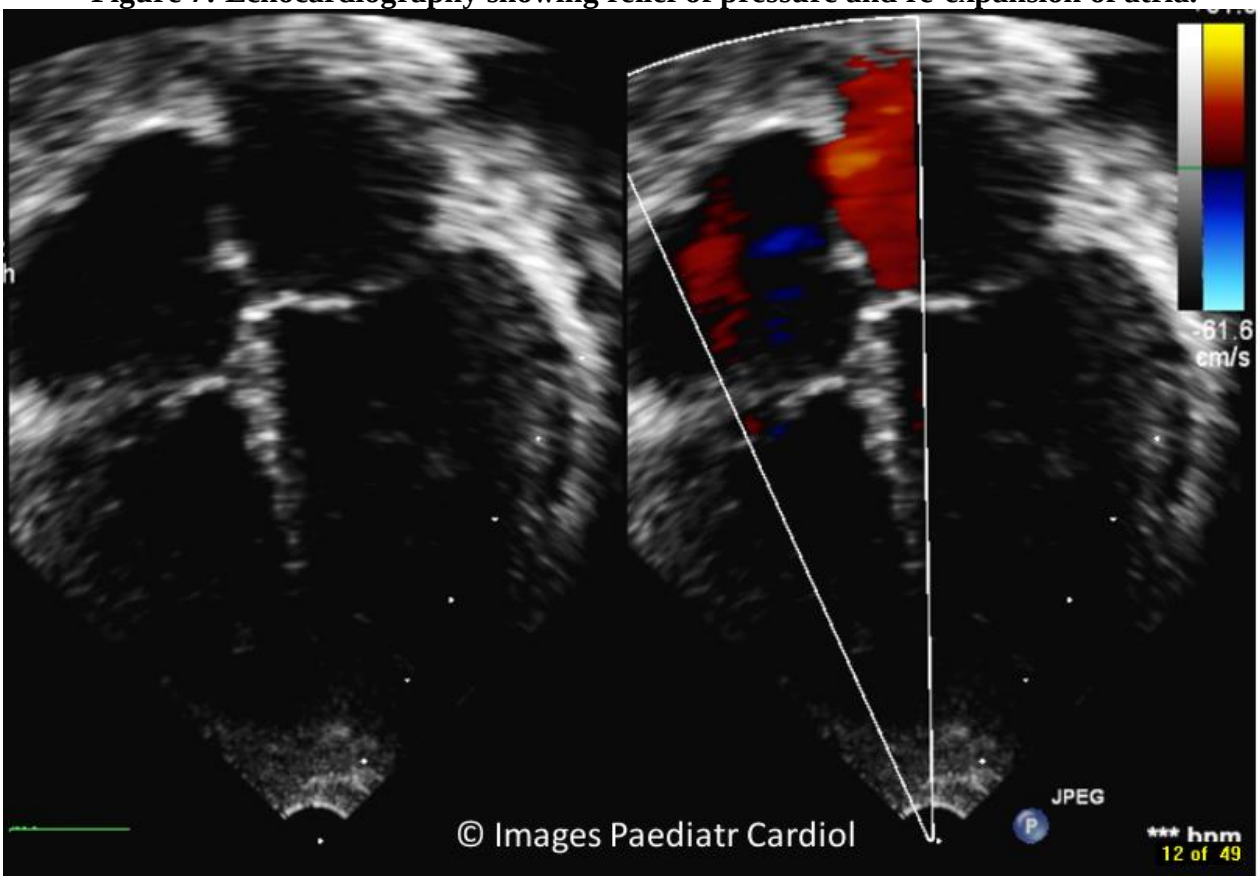
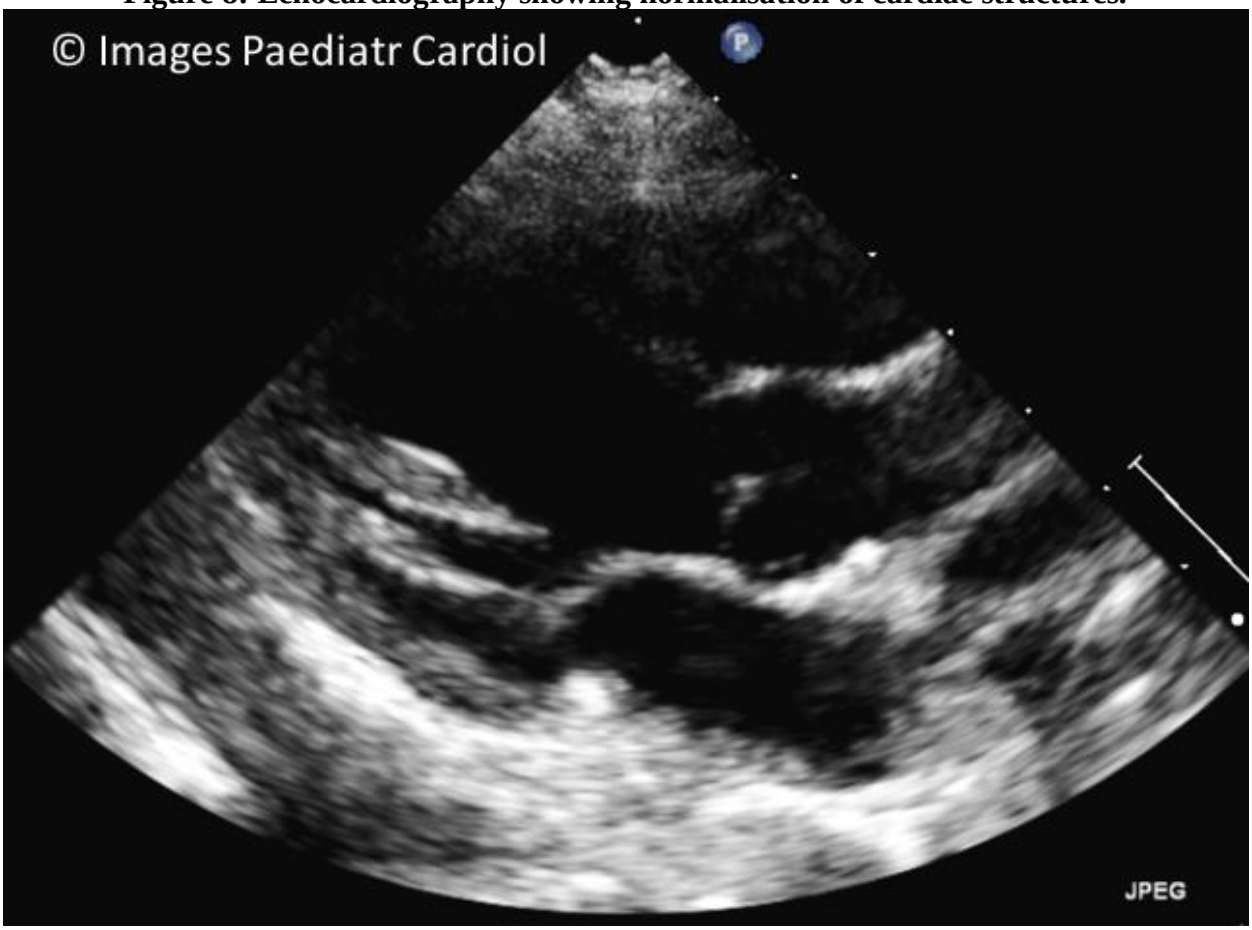


Figure 8: Echocardiography showing normalisation of cardiac structures.



Our patient underwent an uneventful nephrectomy, histology being compatible with Stage I intermediate risk regressive nephroblastoma. He continues to receive post-operative chemotherapy as per the Clinical Management Guidelines Wilms Tumour by the Children's Cancer and Leukaemia Cancer Group.

Discussion

Wilms tumour is the second most common intrabdominal tumour in childhood accounting for approximately 6% of all paediatric cancers. Around 75% of cases occur in children aged less than five years.[1] Children typically present after an incidental discovery of an asymptomatic abdominal mass. Associated signs and symptoms include haematuria, pain, malaise and hypertension.[1] The incidence of haematogenous metastases is approximately 12% and the primary distant site for metastases is the lung.[2,3] Intravenous tumour extension occurs in about 11% of cases with continued extension to the inferior vena cava in about 6% of cases.[1]

Initial radiological investigations include an abdominal ultrasound and chest x-ray. This is followed by MRI of the abdomen and chest, and computerised tomography in order to better stage and anatomically define the primary tumour and assess for distant metastases. Echocardiography is mandatory for patients receiving treatment with doxorubicin and may also be useful in detection of intracardiac tumour extension through the inferior vena cava.[1,6]

Documentation of cardiac function at baseline is important and assessment needs to be continued during treatment, as any deterioration may impact on the administration of anthracycline treatment. Anthracycline-induced cardiotoxicity is a significant cause of morbidity and premature mortality in survivors of childhood cancer, manifesting as a decrease in cardiac function which can lead to congestive heart failure. Up to 60% of patients have been reported to develop echocardiographic abnormality following administration of anthracycline chemotherapy. Echocardiograms are the most commonly used imaging modality in screening for cardiac disease and evaluating cardiac function during and after therapy.[4,5]

In this case, the mediastinal shift and cardiac displacement did not allow for accurate measurement of cardiac function at baseline since the reliability of m-mode views was altered by the distorted anatomy. This distortion also posed difficulties in reliably confirming proximal venous patency and ruling out the presence of venous tumour thrombus. Additionally, echocardiograms enable assessment of outflow obstruction of the great vessels secondary to potential impingement from the mass as well as enabling assessment of the suitability of vessels for central line insertion through Doppler assessment. Echocardiography offers dynamic information with regard to blood flow. However, in this case, there were limitations with regards to confident assessment of the these parameters.

As can be seen from the accompanying images, following initiation of chemotherapy, there was a reduction in size of the pleural metastases allowing for resolution of the mediastinal shift and reversal of the distortion in cardiac anatomy. This enabled more accurate assessment of the venous channels, anatomy and cardiac function.

Conclusion

Distortion of cardiac anatomy presents a diagnostic challenge in accurate assessment of cardiac dynamics and function. In this case, the distortion was caused by pleural metastases from a left nephroblastoma and initiation of treatment with the appropriate chemotherapy protocol resulted in normalisation of the altered anatomy. The initial challenge in assessing venous patency and flow through echocardiography highlights the importance of multimodal imaging including CT and MRI. Also highlighted is the importance of reassessment and monitoring response to therapy which in our

case, translated into improved ability to document parameters of cardiac function following institution of chemotherapy.

References

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