He was seen by our Paediatric team at 9 months old and found to have massive splenomegaly, severe anaemia, neutropenia, lymphopenia and thrombocytopenia. Discussed with Paediatric Haematology who felt this could be a red cell membrane disorder with intercurrent infection and started him on IV co-amoxiclav and folic acid. His fever settled and he was discharged with follow up under the Paediatric Haematology team. Over next 5 months had 6 admissions to Paediatrics with fever and anaemia (see figure 1 for timeline). He required 5 red cell transfusions and was treated with IV antibiotics multiple times. Of note, he was persistently lymphopenic and variably pancytopenic throughout, but his extreme anaemia (lowest Hb 45) was often the focus of his presentations. Between blood transfusions he was very happy and playful, but he was noted to often have abdominal distension following transfusions. He also continued to have a persistent wet cough. He was extensively investigated, including bone marrow aspirate, which were all normal. He was referred to Paediatric Immunology and Genetics, who recommended Whole Exome Sequencing.

On his final admission prior to diagnosis, he presented with fever, pallor and lethargy. He was found to be anaemic again (Hb 71) and given his 5th red cell transfusion however this was unable to be completed due to fever mid-transfusion. He was treated with ceftriaxone as fever of unknown origin. 4 days later he developed abdominal tenderness and massive distension. No clear source of infection could be found. He was discussed with the Haematology team, who accepted him for transfer to their tertiary centre for further investigation and management. They treated him with IV tazocin until fevers settled and he was discharged.

Whole exome sequencing revealed a mutation in the PIK3CD gene, giving him the underlying diagnosis of APDS1 (activated phosphoinositide-3 kinase delta syndrome) or immunodeficiency type 14. There are around 200 known cases of this worldwide with multiple novel APDS mutations. He has now undergone bone marrow transplant and continues under the care of Haematology and Immunology.

Conclusion While primary immunodeficiencies are rarer than haematological causes for pancytopenia and splenomegaly, they must be considered where another cause has not been found. Additionally, whole exome sequencing can provide valuable insights where there is no clear underlying diagnosis for medically significant findings. As general paediatricians we must ensure we advocate for these investigations where necessary.

### Abstract 1032 Figure 1

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### 1083 ALL SLEEP AND NO PLAY? AN AUDIT AND SERVICE EVALUATION OF CHILDREN UNDERGOING RADIOLOGICAL IMAGING

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Aims The use of sedation in children for radiological imaging is common practice in Paediatrics. However, the risks need to be weighed against the benefits of imaging under sedation. Play therapy has been considered as an alternative to sedation in a cooperative child.

This study explored the safety, efficacy and adherence of practice to local trust guidelines for sedation of children (derived from the NICE Sedation under 19s guidelines) and highlighted play therapy as a potential alternative for selected children requiring radiological imaging.

Methods Data was gathered retrospectively from a 6 month period with the help of the Trust’s medical records department. There were 36 children who underwent sedation for various imaging modalities and 19 children who had imaging done utilising play therapy over the same period.

The information gathered from the resources used was collated in an excel database for the purpose of comparative analysis.

Results 1. The assignment of patients was based on their clinical presentation, urgency and medical background

2. Children receiving sedation were predominantly below the age of 3 years while those in the play group were between 6-9 years

3. The youngest child to receive sedation was 3 months old and the youngest to have successful MRI using play therapy was 3 years 5 months

4. The success rate of Sedation was 92% vs. 86% for play therapy

5. 83% underwent MRI, 11% DMSA and 6% MAG3 under sedation. 95% had MRI and 5% CT in the play group

6. 14% required a repeat dose of medication for sedation

Abstracts

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7. None had complications secondary to sedation
8. One had MRI Head done under sedation and later MRI Spine successfully under play therapy at 3 years 5 months
9. Where all documents were available for analysis, the adherence to local guidelines for sedation was 100%

Conclusion
1. Sedation is a safe and effective option available in a DGH setting for young children needing relatively urgent radiological imaging to establish diagnosis where the benefits generally outweigh the risks.
2. Play therapy is a suitable alternative for cooperative children who can be adequately prepared.
3. Healthcare teams and parents need to be made more aware of these options in the future.