recently with hypophosphatasia who is receiving pioneering enzyme replacement treatment.

Baby I was born at 34/40 and required ventilatory support from birth due to respiratory insufficiency. She was noted to have short limbs, hypotonia, and thin ribs on x-rays. Her serum alkaline phosphatase was low; urinary phosphoethanolamine and serum calcium were elevated confirming hypophosphatasia.

In a recent multinational study of 11 patients with severe hypophosphatasia, treatment with recombinant human bone targeted TNSALP (ENB 0040) has been shown to improve bone mineralization. This was associated with healing of rickets, improved developmental milestones and pulmonary function.

Under guidance from the regional Metabolic Bone team at Manchester and with parental consent, Baby I was commenced on ENB 0040 (Asfotase alfa) at the age of 4 weeks with subcutaneous injections three times a week. The drug is being offered to this infant on compassionate grounds by the manufacturer (Alexion pharmaceuticals).

Within 6 weeks of treatment calcium requirement of infant has increased and X-rays have demonstrated remarkable improvement in mineralisation. She remains ventilator dependant with a tracheostomy in situ but, we anticipate that with ENB 0040 treatment, improvement in bone mineralisation and muscle function will facilitate weaning from ventilation.

PF.61 STARRY SKY PATTERN OF FETAL LIVER ASSOCIATED WITH TWIN ANAEMIA POLYCYTHAEMIA SEQUENCE IN MONOCHORIONIC TWINS

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Twin anaemia-polycythaemia sequence (TAPS) is an atypical form of twin-twin transfusion syndrome (TTTS) that presents as a large inter-twin haemoglobin difference with one twin developing anaemia and the other developing polycythaemia, without oligohydramnios-polyhydramnios sequence. The prenatal diagnostic criteria for TAPS require that the middle cerebral artery-peak systolic velocity (MCA-PSV) measure greater than 1.5 multiples of median (MoM) in the anaemic twin and less than 0.8 MoM in the polycythaemic twin.

Starry-sky liver appearance was first described in 1980 by Kurtz *et al,* as corresponding to oedematous swelling of hepatocytes causing decreased echogenicity of the parenchyma (sky) as well as better visualisation of the fibrous walls of the portal vein (stars).

We present a case of monochorionic diamniotic twins with spontaneous twin anaemia- polycythaemia sequence (TAPS) in which the rare sonographic appearance of starry sky liver was seen in the recipient (polycythaemic) twin. The polycythaemic twin had starry sky liver appearance from 25 weeks gestation prior to any sonographic features of TAPS. She was delivered at 28 weeks 6 days gestation when diagnosis of TAPS was made, due to deterioration in growth and Dopplers. One twin was anaemic with haemoglobin of 10.5 g/dl and the other polycythaemic with haemoglobin of 28.0 g/dl.

Conclusion In our case, starry sky appearance of the liver was the early sonographic feature of Twin anaemia polycythaemia sequence.

PF.62 COMPUTATIONAL RECONSTRUCTION OF HUMAN FETAL CARDIAC VENTRICULAR WALL DEVELOPMENT

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We construct a computational model of the electrophysiology of the developing heart.

Anisotropic geometry at ~100 μm voxel resolution was obtained from Diffusion Tensor and Fast Low Angle Shot Magnetic Resonance Imaging and electrical activity from fetal electrocardiograms obtained longitudinally from one gestation and from multiple studies.

Transmural myofibre organisation is established by 136 days gestational age (DGA) and during 2nd and 3rd trimester QR intervals decrease by 20 ms while heart dimensions increase by a 2-fold. This implies an increase in the ventricular conduction velocity. A computational model of the 140 DGA human ventricle is presented that combines cell electrophysiology with anisotropic geometry.

PF.63 A RARE CASE OF FETAL ANAEMIA DUE TO CONGENITAL PYROPOIKILOCYTOSIS TREATED BY INTRAUTERINE FETAL BLOOD TRANSFUSION

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We present the first case of a pre-natal diagnosis of fetal anaemia due to congenital pyropoikilocytosis treated with intrauterine fetal blood transfusion.

A 31 year old woman of Caucasian origin was referred to the fetal medicine unit at 29 weeks gestation with suspected fetal anaemia. She had 3 previous miscarriages and delivered a term female infant who was severely jaundiced and anaemic at birth, requiring multiple exchange transfusions. That infant was diagnosed with congenital pyropoikilocytosis.

In this pregnancy, a middle cerebral artery Doppler peak systolic velocity (MCA PSV) performed at 26 and 28 weeks gestation suggested mild to moderate fetal anaemia. At 29 weeks, MCA PSV indicated severe fetal anaemia. There were no signs of hydrops fetalis. Fetal blood sampling confirmed fetal anaemia and fetal blood transfusion was performed. Fetal blood film confirmed congenital pyropoikilocytosis. At 31 weeks, a repeat fetal blood transfusion was indicated but was unsuccessful due to transient fetal bradycardia. Delivery was prompted and at 32 weeks, a female infant was delivered by elective caesarean section. The infant was anaemic requiring multiple exchange transfusions. Neonatal recovery was uneventful.

Congenital pyropoikilocytosis is an autosomal recessive rare hemolytic anaemia due to an erythocyte membrane defect. It is more often seen in black populations and has rarely been seen in white European populations. Doppler prediction of fetal anaemia using MCA PSV should be advocated in women whose previous pregnancies show them to be at high risk of recurrent fetal or neonatal hemolytic anaemias due to rare erythrocyte defects.

PF.64 COMPLICATED SEQUELAE OF PARVOVIRUS AFFECTED PREGNANCIES

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During the recent epidemic of Parvovirus infection, three complicated pregnancies were managed in the Rotunda Hospital. The fetuses were significantly affected in all three cases, presenting with ultrasonographic findings consistent with severe anaemia; all required intra-uterine fetal transfusions.

Case 1: The first case involved a 30 year old multip who presented at 20 weeks with severe fetal hydrops and a history of Parvovirus