Conclusion: Different IMT cut-off values for each artery are necessary to establish a correct US diagnosis of GCA. These proposed IMT cut-off values may help to improve the diagnostic accuracy of US in clinical practice.

Disclosure of Interests: None declared.


AB0595
INFANTILE TAKAYASU: CLINICAL FEATURES AND LONG TERM OUTCOME

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Background: Takayasu arteritis (TA) is a large vessel vasculitis rarely reported in children, and its incidence is extremely low in infants. Most articles on pediatric TA have not focused on infants. We present the largest case series of infantile TA aiming to characterize demographic and clinical data and compare it with existing data on older children.

Objectives: Characterize demographic and clinical data regarding TA and compare it with existing data on older children

Methods: We conducted an international multi-center retrospective cohort study. Epidemiological and clinical data were collected from patient charts by doctors from six centers.

Results: Twelve patients (50% female) meeting the ACR criteria of TA were included. Median age of symptom onset was 11 months, with a diagnostic delay of 4 months and median time of follow up of 7.5 years. The most common symptoms at presentation were hypertension, BP difference between upper and lower limbs, and fever. The arteries most commonly involved at diagnosis were the abdominal aorta, renal artery, and superior mesenteric artery. Different medications used included steroids, conventional and biological DMARDs, and other immunosuppressive therapies. Half of the patients received biologic agents of which infliximab had the highest complete remission rate (40%). Other medications resulting in complete remission were cyclophosphamide (40%) and methotrexate (38%). Invasive procedures were needed in 58% of patients. The most common complications were cardiac (50%), strokes (42%) and serious infections (33%). None of the patients died.

Conclusion: This study presents the largest series of infantile TA. Compared to reported series on older children, infants with TA were more likely to receive biologic agents, develop complications and require invasive interventions.

Disclosure of Interests: None declared.


AB0596
AORTIC MANIFESTATIONS IN GIANT CELL ARTERITIS: SINGLE CENTRE 10-YEAR EXPERIENCE

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Background: Disease stratification in GCA is an urgent need, with patients categorised into cranial and large-vessel GCA (LV-GCA) subgroups. LV-GCA may have worse outcomes for older children regarding relapsing disease, poor response to glucocorticoids (GC) and aortic involvement.

Objectives: We report a single centre experience using clinical, imaging and treatment outcomes from a specialist clinic.

Methods: 134 patients with LV-GCA were identified over a 10-year period at Southend University Hospital (2012-2022). Medical records were reviewed retrospectively for baseline demographics, clinical presentation, inflammatory markers, imaging (vascular ultrasound, PET-CT, echocardiography), vascular damage and treatment.

Results: There was a female predominance (female:male=91:9). Age at presentation ranged from 46 to 86 years (median 70 years). Co-morbidities implicated in aortic disease included hypertension (n=60), hypercholesterolaemia (n=29), diabetes (n=14), aortic valve disease (n=5) and other aortic diseases including coronary and carotid artery disease (n=19). Constitutional disturbance was most frequently observed presentation (70%, n=94), and the only feature for 11 patients. This was followed by cranial symptoms (62%, n=83), polymyalgia (53%, n=71), ischaemic symptoms i.e., visual disturbance or tongue/jaw claudication (24%, n=32) and cardiovascular presentations (7%, n=9). The latter included limb claudication, stroke, and aortic aneurysm. Although LV-GCA refers to extra-cranial disease, 12 patients (9%) had isolated cranial and/or ischaemic symptoms at initial presentation.

Inflammatory markers were typically elevated at presentation, C-reactive protein ranged from 1-425mg/L and ESR 1-130mm. Vascular ultrasound was used in diagnosis in 93 patients, with positive temporal arterial findings in 50% (n=38) and positive axillary findings in 75% (n=57). PET-CT data was available for 125 patients, of which 113 were positive for LV-GCA. The positive arterial uptake was seen in 77%, with 7 ascending and 1 abdominal aortic aneurysm observed. Transthoracic echocardiogram was available for 46% (n=62). Four (6.5%) patients had a dilated aortic root when indexed to height as per British Society of Echo-cardiography (BSE) guidelines[1] (SOV (mm/m²) > 21.8mm in males and >20.7mm in females). Values for our patients were 22.6 and 21.2 mm/m² for the female patients and 29.2 and 25.2 mm/m² for the male patients. Furthermore, 32 patients showed some extent of diastolic dysfunction as per BSE criteria (52%). All patients received GC as part of their treatment, 60% (n=82) needing one or more DMARDs and 17% (n=23) tocilizumab for relapsing disease. DMARDs used included Leflunomide (n=63), Methotrexate (n=16), Mycophenolate mofetil (n=3) and Azathioprine (n=1). One patient received cyclophosphamide.

Conclusion: By combined imaging modalities, 11 patients (8%) had evidence of ascending aortic damage. Grade 1 diastolic dysfunction can be age related, so this may be association rather than causation[2]. Over half of patients had not undergone echocardiogram evaluation, so there may be a hidden burden of disease. Many patients required GC-sparing therapy, showing GC alone are often not enough to halt disease progression, and vascular damage was relatively reduced compared to historical reports. The authors feel GCA services should include standardised protocols for early DMARDs, continuing thorough assessment for LV-GCA and vascular damage, including echocardiography, progressing to cross-sectional imaging if indicated.

REFERENCES:

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AB0597
FDG-PET/CT IN THE DIAGNOSE AND FOLLOW UP OF TAKAYASU VASCULITIS


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Background: Takayasu vasculitis (TAK) is a chronic disease, where clinic and serological markers as CRP/ESR may fail to predict development of new vascular lesions in the disease course[1]. Similarly, [2]fluorodeoxyglucose positron emission tomography/computed tomography (FDG-PET/CT) studies show conflicting results on the association between vessel uptake of FDG and clinical and laboratory finding. A study on new FDG-PET activity scoring system, PETVAS was newly published but has not been validated in other cohorts[2]. To date there are limited data on FDG-PET/CT finding at time of diagnoses before treatment induction and 18-FDG uptake and development of new stenosis during follow-up[3].

Objectives: The goal of this study was to see: 1) FDG-PET/CT uptake in newly diagnosed patients before any treatment start 2) FDG-PET/CT uptake and development of new vascular lesions during follow up magnetic resonance angiography (MRA) 3) assess PETVAS score before and after treatment induction.

Methods: All patients in a population-based TAK cohort with FDG-PET/CT at the time of diagnoses and treatment induction were identified. Disease activity was assessed with the NIH activity score[1]. Patients that had clinical, laboratory and MR-angiography prior to/or right after FDG-PET/CT and a minimum of one follow up MRA. The clinical report from the FDG-PET/CT and MRA were reviewed and arteries/aorta regions of interest (supraaortic arteries, aorta, iliac and femoral arteries) scored from 0-3, where 0 represent no uptake, 1 less then liver, 2 same as liver and 3 higher than liver and finally summarised these to PETVAS score.