scored using the JDM severity score tool. The histo-pathologists' overall severity score (VAS) varied from to 1 to 9/10; the highest scores were observed in the two patients who died.
Conclusion: In the present series of patients with JDM, TIF-1-γ Ab-associated-clinical phenotypes and ethnicities are more heterogeneous than previously reported. TIF-1-γ Ab is associated with a high mortality rate in a subset of patients.

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AB0936 A NATIONAL SURVEY OF CLINICAL PRACTICE OF CORTICOSTEROID USE IN NEWLY DIAGNOSED OR FLARING CASES OF JUVENILE IDIOPATHIC ARTHRITIS ACROSS THE UK

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Background: Corticosteroids (CS) are widely used for rapid-action or induction treatment in children and young people (CYP) with juvenile idiopathic arthritis (JIA). Given a lack of evidence base on CS induction regimen for CYP with JIA, and since criteria for choosing CS are based on healthcare professional (HCP) preference, further research is needed (1).
Objectives: To establish the opinions of HCPs current practice regarding the clinical criteria for commencing CS treatment
Methods: A national electronic survey was undertaken among HCPs across the UK as part of the Steroid Induction Regimen for Juvenile Idiopathic Arthritis (SIRJIA) study.
Results: A total of 39 (24%) responses were received from 162 HCPs. These included 22 (56%) NHS consultants, five (13%) grid trainees, eight (21%) clinical nurse specialists and four other HCPs (10%).
The most common treatments in CYPs with newly diagnosed JIA or a disease flare were intra-articular (IA) CS or a combination of DMARDs and IAS (except for systemic JIA and oligoarticular JIA). The majority of HCPs 17 would treat new and flaring CYP the same in terms of a CS remission induction regime, with 53% choosing a different regime or not answering.
The key criteria that HCPs used for commencing CS and choosing the route of administration were rapid induction of remission (31 (89%)), high disease activity (31 (89%)), severity of systemic JIA (30 (86%)) and level of inflammation (28 (80%)) Table 1. The number one determinant of route of administration of CS was disease severity followed by disease subtype.
The majority of HCPs (52-72% depending on role) would consider entering CYP with JIA into a trial randomising to the various modes of administration.

Table 1:

Reasons of CS Choice	Number N=39	Percentage%
High Disease Activity	35	89.7
Rapid induction of remission	34	87.2
Severity of Systemic JIA	34	87.2
Level of inflammation	32	82.5
Severe Uveitis	30	76.9
JIA subtype	27	68.2
Targeting Specific Joints	26	66.7
Level of Disability	18	46.2
Level of pain	16	41.0
Long-standing Disease	11	28.1
Patient reluctance to take DMARDS	8	20.5

Conclusion: The results from this national survey of clinical practice showed varying practices in the management of new CYP with JIA and those that are flaring. The majority of HCPs who completed this survey, indicated that they would be prepared to consider entering CYP into a trial that randomised to the four CS delivery methods.

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AB0937 IDENTIFYING THE PRIMARY OUTCOME MEASURE AND PROTOCOL COMPONENTS FOR A PROSPECTIVE FEASIBILITY STUDY OF CORTICOSTEROID REGIMENS FOR CHILDREN AND YOUNG PEOPLE WITH JUVENILE IDIOPATHIC ARTHRITIS USING CONSENSUS METHODS WITH YOUNG PEOPLE, FAMILIES AND PROFESSIONALS

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Background: Juvenile idiopathic arthritis (JIA) is an umbrella term for seven relapsing-remitting inflammatory conditions in children and young people (CYP). Early, intensive treatment can prevent long-term damage; however, established drugs exhibit a delayed response, prompting the need for rapid-onset treatment in the form of corticosteroids. Given a lack of consensus as to which corticosteroid induction regimen should be used for CYP with JIA, a feasibility trial of different regimens is needed.

Objectives: The aim was to achieve consensus among CYP, families, and healthcare professionals (HCPs) about the primary outcome measures and protocol components to include in a prospective feasibility study.
Methods: A modified Nominal Group Technique was used to achieve consensus on the most appropriate primary outcome measure to be included in a prospective feasibility study, in addition to other protocol components such as inclusion/exclusion criteria. Fifteen participants participated in the process, including a combination of CYP with JIA, families (n=9) and HCPs (n=6).
Results: In the first vote, participants agreed that 'Juvenile Arthritis Disease Activity Score (JADAS)' and 'Physician Global Assessment Score' were most meaningful. During sub-group discussions, the need for a composite score which captured the voice of CYP and families was emphasised. In the second vote, 'JADAS' and the 'JIA Core Set' were identified as the most important. Further discussions led to the results of the third vote, agreeing 'JADAS' as the primary outcome measure of choice being measured at 6 weeks after commencement of treatment. The majority of HCPs, CYP and families voted for all JIA sub-types to be included in a prospective feasibility study, with some queries about the inclusion of systemic JIA given its unique presentation. Participants also identified the need for more frequent data collection time points to capture the rapid onset of corticosteroid action, while CYP and families opted for accessible mechanisms for participation, such as digital follow-up strategies.
Conclusion: It is feasible to include CYP, families and HCPs in synthesising complex concepts to agree by consensus the design components of clinical research. The primary outcome measure for inclusion in a prospective feasibility study of corticosteroid regimens in CYP with JIA was co-prioritised, with CYP and families taking a leading role in the ultimate selection of an appropriate outcome measure and other study protocol components. Using consensus methods with CYP, families and HCPs is a systematic and rigorous way in which to select outcome measures that are both meaningful and relevant to everyone involved in the care and treatment of CYP with JIA.

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Disclosure of Interests: Simon Stones Consultant for: SS has provided consultancy services to Envision Pharma Group, though this is not related to the contents of this abstract., Speakers bureau: SS has undertaken speaking engagements for Actelion, eyeforpharma, Four Health, Janssen and Roche, though these are not related to the contents of this abstract., Heather Bagley: None declared, Ashley Jones: None declared, Flora McErlane: None declared, Tracy Moitt: None declared, Gloria Nkhoma: None declared, Frances Sherratt: None declared, Bridget Young: None declared, Michael Beresford: None declared, Eileen Baildam Consultant for: EB has received one paid consultancy for Pfizer in the last 12 months, but this was not related to this study.
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AB0938

EFFICACY AND SAFETY OF BIOLOGICAL THERAPY WITH ETANERCEPT IN A CASE OF SEVERE POLIARTHRITIS ASSOCIATED TO HARLEQUIN ICHTIOSIS

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Background: Harlequin Ichthiosis (HI) is a rare autosomical recessive congenital disease, due to mutations of gene ABCA12.The estimated incidence is approximately 1 in 300,000 births, and approximately 200 cases have been reported worldwide. Typical manifestations of the disease at birth are the presence of hyperkeratotic plates with erythematous fissures, ectropion (eversion of the lower eyelids) and eclabium (eversion of the lips), rudimentary ears and nasal hypoplasia, and articular contractures. Babies who survive into infancy tend to loose hyperkeratotic plaques in favour of generalized scaling and erythroderma. Mental retardation is present in about 2/3 of cases.The association between HI and inflammatory joint involvement has been reported only in few patients since now.
Objectives: We are describing the case of a 6 years old kid with HI that at the age of 4 developed arthritis with severe impact on his quality of life.
Methods: We report clinical and laboratory findings, treatment choices and outcomes of young boy with HI who developed polyarthritis.
Results: A 6 years old kid with HI (the genetic test showed homozigous mutation of ABCA12) came to our attention with an history of chronic arthritis.
Since he was 4, he developed bilateral knee arthritis. At first, he was treated with antimicrobial drugs, without improvement, in suspect of septic arthritis. In the following two years, he developed severe chronic poliarthritis.At the first visit in our center, he showed the classical clinical feature of HI (severe ectropion, contracture and generalized erythroderma), and had history of multiple severe infections and sepsis. Arthritis affected both hips, knees, ankles, wrists, elbows, shoulders and all the feet joints. He lost the ability to walk since the onset of arthritis.
Laboratorial features showed negative anti nuclear antibodies (ANA) and anti. extractable nuclear antigen antibodies (ENA), anti ciclic citrullinated peptide and rheumatoid factor. No signs of uveitis were present at ophtalmological evaluation.
The patient was treated with intraarticular corticosteroid injections (IACI) into knees, wrists, elbows and ankles, and then oral methotrexate (MTX) was started.
The clinical response was initially good, with partial recovery of deambulation in the 4 months following the intrarticular injection treatment. After 7 months ankles and wrists showed swelling and tenderness, and IACI were repeated, with clinical improvement.

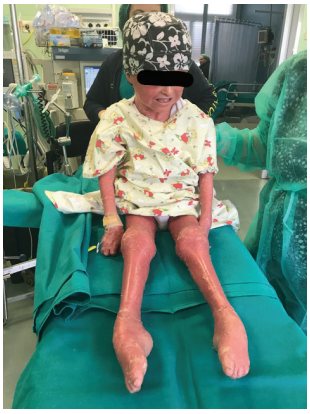

Figure 1

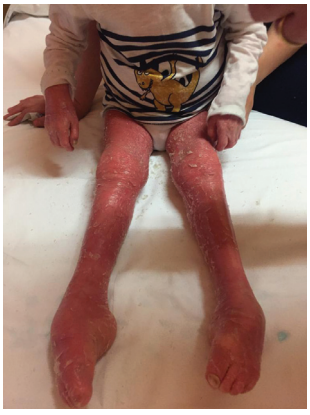

Figure 1