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2020 ACR Guideline for the Treatment of JIA: Therapeutic Approaches for Oligoarthritis, TMJ Arthritis, and Systemic JIA, Medication Monitoring, Immunizations and Non-Pharmacologic Therapies

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NOVEMBER 5-9
#ACR20

COI DISCLOSURE INFORMATION

Karen Onel

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I have no relevant financial relationships to disclose

HSS

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Please Note:

This presentation contains *DRAFT* recommendations that are pending full approval from the American College of Rheumatology

Recommendations—Not all will be presented today

Topic	Recommendations	Strength of Recommendation		Quality of Supporting Evidence			
		Conditional	Strong	Very Low	Low	Moderate	High
Oligoarthritis	8	4	4	6	2	0	0
TMJ Arthritis	5	4	1	5	0	0	0
Systemic JIA	9	5	4	9	0	0	0
Medication Monitoring	15	14	1	15	1	0	0
Infections/Immunization	6	3	3	5	0	0	0
Non-Pharmacologic Rx	2	1	1	2	0	0	0
Imaging	2	1	1	2	0	0	0
Total	47	32	15	44	3	0	0

KEY

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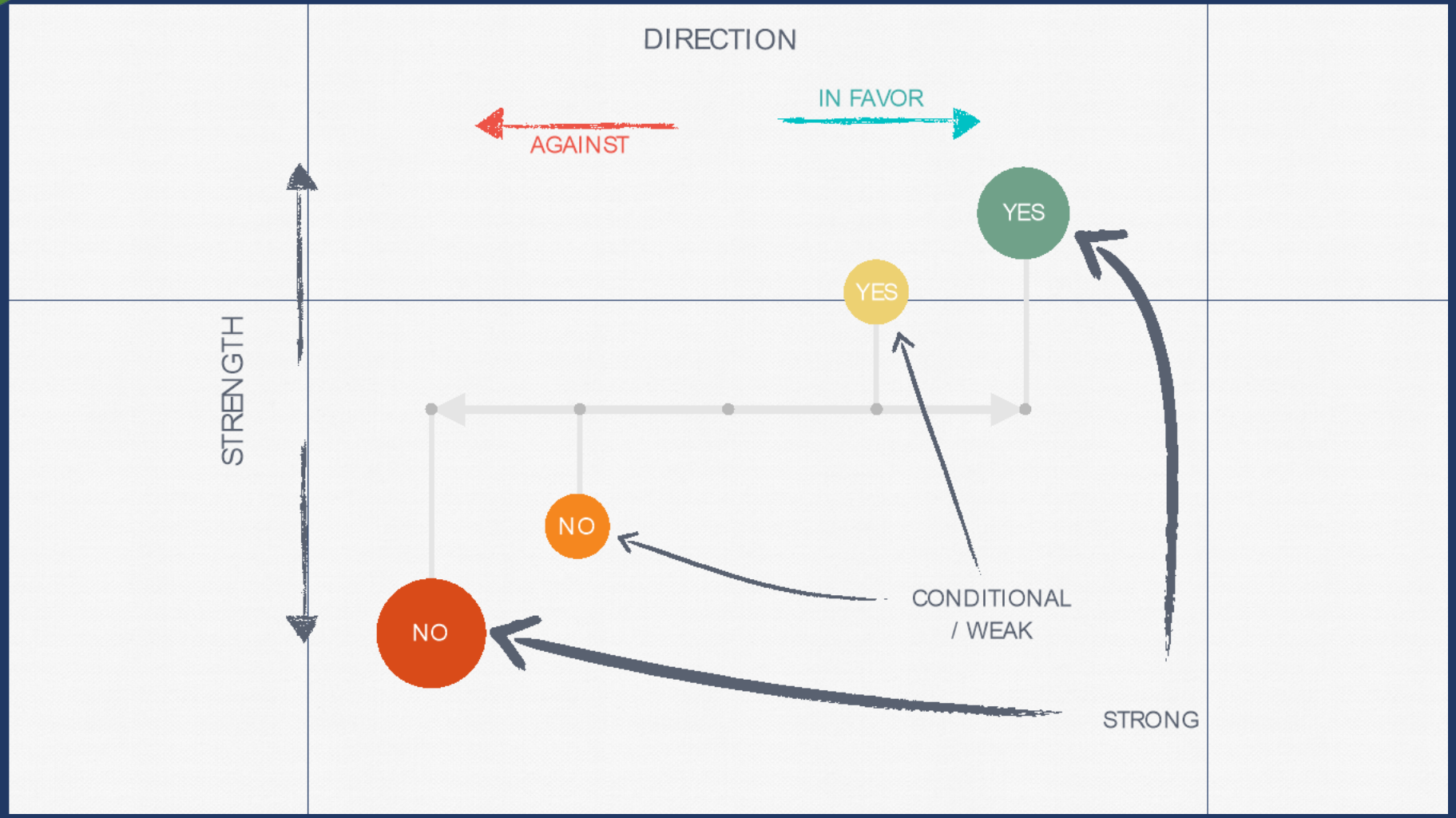
- **GRADE methodology**
  - Certainty in the evidence = ‘quality’
    - high, moderate
    - low L or very low VL
  - Strength of recommendations
    - Strong S S or conditional C C
    - Depends on quality of evidence
      - Patient preferences and values
      - Weighing benefits and harms

www.gradeworkinggroup.org

Strength of recommendations



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The following recommendations are intended for children in whom the diagnosis of JIA has been already been made

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Oligoarthritis: < 5 involved joints

- Intraarticular glucocorticoids (IAGC) are recommended as part of initial therapy **S** VL
  - THA is recommended as preferred agent **S** L
- A trial of consistent NSAIDs is recommended as part of initial therapy **C** VL
- Oral glucocorticoids should not be added to initial therapy **C** VL

- Non-biologic DMARDs are recommended if no or incomplete response or intolerance to NSAIDs and/or IAGC S VL
  - Methotrexate is recommended as preferred agent C L
- Biologic DMARDs are recommended if no or incomplete response or intolerance to NSAIDs and/or IAGC and at least one non-biologic DMARD S VL
  - There is no preferred agent

- Risk factors (e.g., involvement of ankle, wrist, hip and/or TMJ, presence of erosive disease, delay in diagnosis, elevated inflammatory markers, symmetric disease) are recommended to guide treatment decisions C VL
- Validated disease activity measures are recommended to guide treatment decisions C VL
- Imaging guidance is recommended for IAGC of joints that are difficult to access or to specifically to localize the distribution of inflammation C VL

TMJ arthritis: may or may not be isolated

- Intraarticular glucocorticoids (IAGC) are recommended as part of initial therapy C VL
  - There is no preferred agent
- A trial of consistent NSAIDs is recommended as part of initial therapy C VL
- Oral glucocorticoids should not be added to initial therapy C VL

- Non-biologic DMARDs are recommended if no or an incomplete response or intolerance to NSAIDs and/or IAGC **S** VL
  - Methotrexate is recommended as preferred agent **C** VL
- Biologic DMARDs are recommended if no or an incomplete response or intolerance to NSAIDs and/or IAGC and at least one non-biologic DMARD **C** VL
  - There is no preferred agent

Systemic JIA (sJIA) with or without Macrophage Activation Syndrome (MAS)

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sJIA without MAS

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- NSAIDs are recommended as initial monotherapy C VL
- Biologic DMARDs (IL-1 and IL-6 inhibitors) are recommended as initial monotherapy C VL
  - There is no preferred agent
- A single biologic DMARD is recommended over a combination of non-biologic therapies if no or incomplete response or intolerance to NSAIDs and at least one non-biologic DMARD S VL

- Oral glucocorticoids should not be used as initial monotherapy

- Non-biologic DMARDs (e.g.methotrexate, cyclosporine, sulfasalazine, leflunomide, thalidomide) should not be used as initial monotherapy

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sJIA with MAS

A quick word about MAS

Major cause of morbidity and mortality for children with sJIA

- Cytokine storm
- Secondary hemophagocytic syndrome
- Can be seen with any rheumatic disease; most commonly seen with sJIA
- Features include fever, high ferritin, cytopenias, elevated lft's, high triglycerides

- Glucocorticoids are recommended as initial monotherapy C VL
- Biologic DMARDs (IL-1 and IL-6 inhibitors) are recommended over calcineurin inhibitors in order to achieve inactive disease and resolution of MAS C VL
  - There is no preferred agent
- Biologic DMARDs and non-biologic DMARDs are recommended over chronic glucocorticoid treatment if there is residual arthritis and an incomplete response to IL-1 or IL-6 inhibitors S VL
  - There is no preferred agent

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sJIA with or without MAS
Inactive Disease

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- Tapering and discontinuing glucocorticoids is recommended after inactive disease state has been attained **S** VL

- Tapering and discontinuing biologic DMARDs is recommended after inactive disease state has been attained **C** VL

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These recommendations pertain to all children with JIA

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Medication Monitoring
Laboratory Testing and Infection Screening

- **<u>Laboratory Monitoring</u>**
  - CBC and LFT's
  - Kidney function as appropriate
- Should be checked prior to treatment
- May be repeated as needed for clinical care

1-2 months Every 3 months thereafter	Every 6 months	Once Yearly	None required
Methotrexate	**NSAIDs**	**Hydroxychloroquine***	**TNFi**
Sulfasalazine			**Abatacept**
Leflunomide			
Tocilizumab*			
Anakinra			
Canakinumab			

*Baseline and annual retinal screening are recommended over starting annual screening 5 years after treatment onset C VL

*Monitoring lipids every 24 weeks after starting Tocilizumab as per package insert is recommended C VL

Infection Monitoring

Controversial

Risk is dependent on who you are and where you live

- TB screening is recommended prior to starting biologic DMARD therapy and when there is a concern for exposure thereafter C VL
- We could not reach consensus on whether to screen for other viral infections prior to DMARD therapy (e.g.Hepatitis B and C) VL

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Immunization

Children with JIA can respond to immunizations with no increased risk of flare
Which immunizations are recommended*:

- No immunosuppressive medications:
  - Immunizations(live and inactivated) are recommended for children with JIA not on immunosuppression S VL
- Immunosuppressive medications
  - Inactivated vaccines are recommended for children with JIA on immunosuppression including PPSV23 S VL
  - Live attenuated vaccines are not recommended for children with JIA on immunosuppression C L

*As per CDC and AAP guidelines, including annual flu shots

Special Circumstances

- Prior to medication for the unimmunized or underimmunized
  - Immunization is recommended for children with active non-systemic JIA with no evidence of immunity to MMRV prior to starting immunosuppressive medications C VL
- Use of live virus vaccines in the household of children receiving immunosuppressive medication
  - Live attenuated vaccines are recommended in the household of children with JIA on immunosuppression as per CDC guidelines S VL

Non-pharmacologic management

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- PT/OT is recommended regardlessof concomitant medical therapy C VL
- A discussion of healthy, age-appropriate diet is recommended S
- Special diets should not be used to specifically treat JIA S VL
- Supplements and/or herbal interventions should not be used to specifically treat JIA C VL
- X-ray should not be used as a screening test prior to advanced imaging for the purpose of identifying active synovitis or enthesitis S VL

- These guidelines demonstrate the evolution of the treatment of JIA
- There is decreased reliance on NSAIDs and Glucocorticoids and earlier introduction of biologic DMARDs
- Although evidence remains very low and many recommendations are conditional, the inclusion of parents and patients in the decision making process strengthens their validity
- Finally, it is important to remember that these are guidelines. Clinical care remains in the hands of the care provider and patient.

Special thanks to the children and families who trust us to make these recommendations and give our work meaning.

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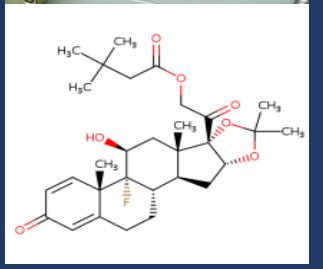
We need this in the US!



Triamcinolone Hexacetonide

Thank You!

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