SYSTEMIC JUVENILE IDIOPATHIC ARTHRITIS

DRUG NAME	BRANDS REIMBURSED	DOSAGE FORM/ STRENGTH	REIMBURSEMENT CRITERIA	STANDARD APPROVAL DURATION
Anakinra	Kineret	100mg/0.67mL pre-filled syringe	**For the treatment of systemic juvenile idiopathic arthritis in patients** who meet the following criteria; • Patient must have a diagnosis of sJIA with fever (>38 degrees Celsius) for at least 2 weeks AND at least ONE of the following: o rash of systemic JIA o serositis (e.g. pericarditis , pleuritis, or peritonitis) o lymphadenopathy (e.g. cervical, axillary, inguinal) o hepatomegaly o splenomegaly • The physician making the request has ruled out other potential etiologies (e.g. malignancies, serious clinical infections, and other inflammatory or connective tissue diseases); AND • Age of disease onset is younger than 16 years of age. (Note: the physician must specify age of disease onset in the request); AND • Systemic corticosteroids cannot be used for at least ONE of the following reasons (please specify name and current dose of corticosteroid, if applicable): o The patient is unresponsive and/or refractory to systemic corticosteroids; OR o The patient has experienced a systemic reaction (e.g.	Initial: 1 year

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Anakinra	Kineret	100mg/0.67mL pre-filled syringe	fever, rash of sJIA, serositis, lymphadenopathy, hepatomegaly or splenomegaly) while on tapering doses of systemic corticosteroids (i.e. the patient is corticosteroid dependent); OR ◦ The patient has experienced an adverse drug reaction to a systemic corticosteroid; OR ◦ The use of systemic corticosteroids is contraindicated in this patient. Note: The following requests will undergo external review on a case-by-case basis: • Patients with Macrophage Activation Syndrome • Patients who meet initial sJIA criteria and are currently 16 years of age or older • Patients who meet initial sJIA criteria and are requesting higher dosing regimens (Please provide rationale for the higher dosing regimen with your request) Dosing: 1-2 mg/kg subcutaneously once daily. **Renewal** will be considered for patients demonstrating at least a 50% reduction in corticosteroid dose (unless contraindicated, not tolerated, unresponsive or refractory at the time of initial request) and no evidence of active systemic disease. For renewals beyond the second year, objective evidence of preservation of treatment effect must be provided. The following renewal requests will undergo external review: • Evidence of active systemic disease • Requests for higher dosing regimens (Please provide	Renewal: 1 year

DRUG NAME	BRANDS REIMBURSED	DOSAGE FORM/ STRENGTH	REIMBURSEMENT CRITERIA	STANDARD APPROVAL DURATION
			rationale for the higher dosing regimen with your request) • Patient is currently 16 years of age or older	
Tocilizumab	Actemra	80 mg / 4 mL 200 mg / 10 mL 400 mg/ 20 mL	**For the treatment of systemic juvenile idiopathic arthritis in patients** who meet the following criteria; • Patient must have a diagnosis of sJIA with fever (>38 degrees Celsius) for at least 2 weeks AND at least ONE of the following: o rash of systemic JIA o serositis (e.g. pericarditis , pleuritis, or peritonitis) o lymphadenopathy (e.g. cervical, axillary, inguinal) o hepatomegaly o splenomegaly • The physician has ruled out other potential etiologies (e.g. malignancies, serious clinical infections, and other inflammatory or connective tissue diseases); AND • Age of disease onset is younger than 16 years of age. (Note: the physician must specify age of disease onset in the request); AND • Systemic corticosteroids cannot be used for at least ONE of the following reasons (please specify name and current dose of corticosteroid, if applicable): o The patient is unresponsive and/or refractory to systemic corticosteroids; OR o The patient has experienced a systemic reaction (e.g. fever, rash of sJIA, serositis, lymphadenopathy, hepatomegaly or splenomegaly) while on tapering doses of systemic cortico-steroids (i.e. the patient is corticosteroid dependent); OR	1 year (Initials and renewals)

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Tocilizumab	Actemra	80 mg / 4 mL 200 mg / 10 mL 400 mg/ 20 mL	◦ The patient has experienced an adverse drug reaction to a systemic corticosteroid; OR ◦ The use of systemic corticosteroids is contraindicated in this patient. Note: The following requests will undergo external review on a case-by-case basis: • Patients with Macrophage Activation Syndrome • Patients who meet initial sJIA criteria and are currently 16 years of age or older • Patients who meet initial sJIA criteria and are requesting higher dosing regimens (Please provide rationale for the higher dosing regimen with your request) Dosing: For those less than 30 kg,12 mg/kg IV every 2 weeks For those greater than or the same as 30 kg 8 mg/kg IV every 2 weeks Note: Recommended maximum adult dose is 800mg. **Renewal** will be considered for patients demonstrating at least a 50% reduction in corticosteroid dose (unless contraindicated, not tolerated, unresponsive or refractory at the time of initial request) and no evidence of active systemic disease. For renewals beyond the second year, objective evidence of preservation of treatment effect must be provided. The following renewal requests will undergo external review: • Evidence of active systemic disease • Requests for higher dosing regimens (Please provide	Renewal: 1 year

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			rationale for the higher dosing regimen with your request) • Patient is currently 16 years of age or older	