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International Pediatric ORL Group (IPOG) laryngomalacia consensus recommendations

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Running head: Laryngomalacia

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Abstract

Objective
To provide recommendations for the routine management of pediatric patients that present with signs concerning for laryngomalacia.

Study Design
Expert opinion by the members of the International Pediatric Otolaryngology Group (IPOG).

Results
Consensus recommendations include evaluation and treatment considerations for commonly debated issues in laryngomalacia, initial work-up of those patients presenting with inspiratory stridor, treatment recommendations based on disease severity, management of the infant with feeding difficulties, post-surgical treatment management recommendations, and suggestions for acid suppression therapy. These recommendations are based on the collective opinion of the IPOG members and are targeted to primary care practitioners, otolaryngologists, and other health care providers who commonly evaluate pediatric patients with noisy breathing.

Conclusion
Laryngomalacia care consensus recommendations are aimed at improving patient-centered care in infants with laryngomalacia.

Level of evidence: 5
Keywords: laryngomalacia, infant, stridor, pediatric

International Pediatric ORL Group (IPOG)

Laryngomalacia Consensus Recommendations

Consensus Objectives
To provide recommendations for the routine management of pediatric patients that present with signs concerning for laryngomalacia.

Target Population
Pediatric patients with signs concerning for laryngomalacia.

Intended Users
These consensus recommendations are targeted for:

1. Primary care practitioners, otolaryngologists, and other health care providers who commonly evaluate pediatric patients with noisy breathing.

Recommendations and Justification
The recommendations are outlined in the following appendices

- ***Section 1:*** Evaluation and treatment considerations
- ***Section 2:*** Initial presentation algorithm
- ***Section 3***: Comprehensive care algorithm
- ***Section 4:*** Management of the difficult to feed infant with laryngomalacia
- ***Section 5:*** Post-surgical treatment algorithm
- ***Section 6:*** Recommendations for acid suppression therapy

Disclaimer
Members of the International Pediatric ORL Group (IPOG) prepared this report. Consensus recommendations are based on the collective opinion of the members of this group. Any person seeking to apply or consult the report is expected to use independent medical judgment in the context of individual patient and institutional circumstances.

Section 1: Evaluation and treatment considerations

The members of the IPOG identified five frequently debated evaluation and treatment considerations in the management of laryngomalacia. Variation in practice among the current group members remains, and the purpose of this section is to provide a list of reasonable options based on expert opinion. **Table 1.**

Table 1.

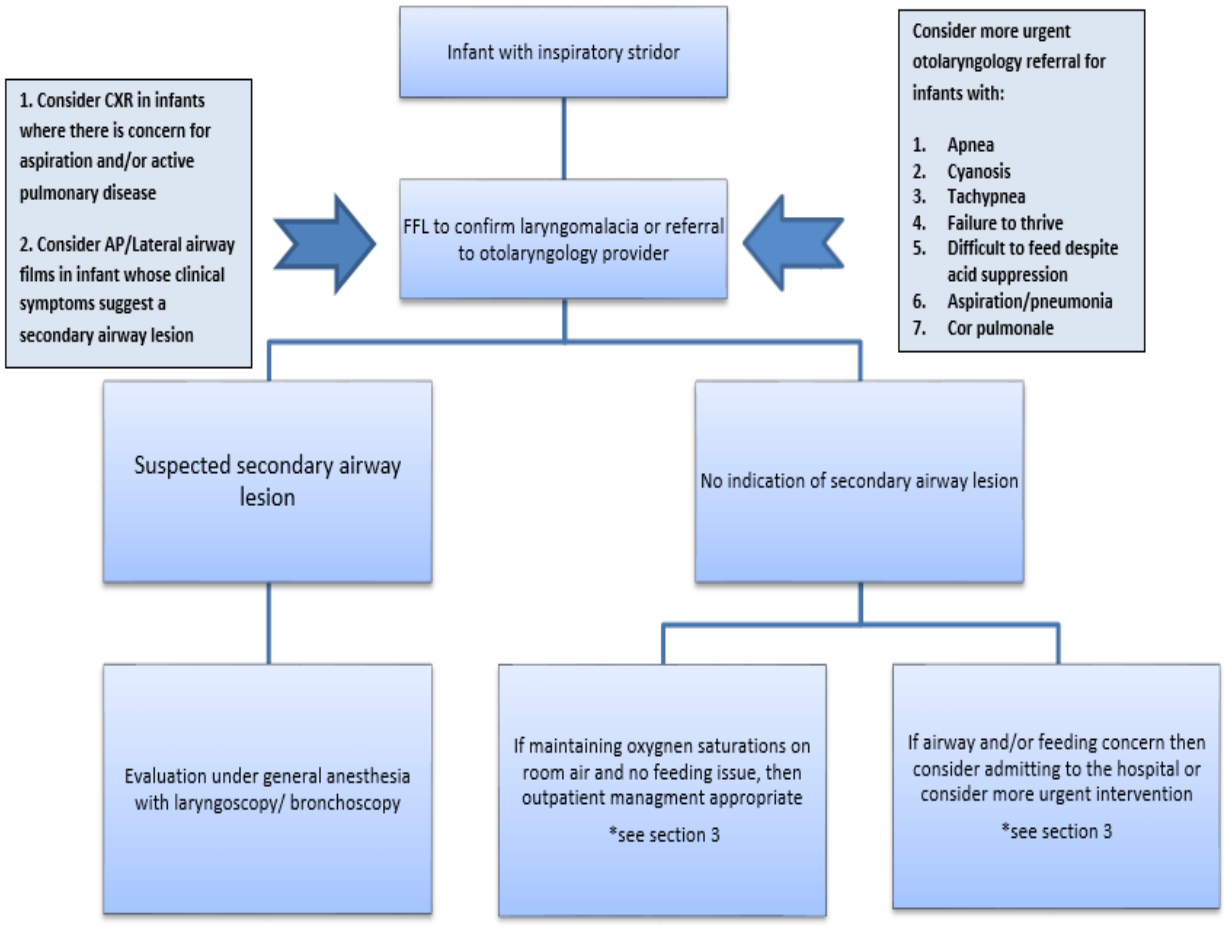
Question	
1. What findings on initial presentation should prompt a more urgent evaluation by an otolaryngologist?	• Apnea • Tachypnea • Cyanosis • Failure to thrive • Difficult to feed despite acid suppression/texture modification • Aspiration/pneumonia • Cor pulmonale
2. Should I treat laryngomalacia empirically with acid suppression?	• Yes, if child having feeding and/or respiratory difficulties • Consider observation in those infants with mild respiratory symptoms and are gaining weight appropriately • Can use either step-up or step-down methodology (**see Section 6**) • Recommend weaning acid suppression based on symptoms vs. stopping abruptly • Consider GI referral for concurrent management
3. Should I formally assess the infant's swallow?	• Consider feeding/swallow evaluation and diet modification in cases where there is cough, choking, regurgitation, feeding difficulty, no weight gain, or failure to thrive • Strongly consider evaluation in children with evidence of aspiration or those with neurologic disease • Consider evaluation by either/both video fluoroscopic swallow study (VFSS) and/or fiberoptic endoscopic evaluation of swallowing (FEES). Assessment in conjunction with feeding therapy may aid diagnostic accuracy and feeding recommendations • Consider acid suppression in patients with laryngeal penetration and/or aspiration on swallow evaluation
4. What other consultations should I consider for the infant with severe disease?	• Pulmonary evaluation if disease on imaging or symptoms of asthma/reactive airway disease • Consider polysomnography or home oximetry monitoring if significant apnea • Cardiac consultation if heart disease suspected • Possible GI evaluation if refractory to acid suppression therapy • Neurology and/or brain MRI if neurologic disease suspected (i.e. physical findings of hypotonia, pooled/frothy secretions on endoscopy) • Genetics evaluation for those with craniofacial dysmorphism or severe disease • Craniofacial team evaluation for those with craniofacial anomalies
5. What assessment should be done for persistent symptoms after supraglottoplasty?	• Suggestions include: ▪ Consider aerodigestive evaluation including pH/impedance probe to rule out persistent reflux, esophageal biopsies to rule out eosinophilic esophagitis, pulmonary evaluation to optimize respiratory function ▪ Consider gastrostomy tube and/or fundoplication for patients with esophageal reflux not managed on maximal medical

	therapy ▪ Consider polysomnography in patients with oxygen desaturations or signs of apnea ▪ Consider neurology and/or MRI brain if neurologic disease suspected ▪ Consider tracheostomy in patients with multiple co-morbidities or synchronous airway lesions

Section 2: Initial presentation algorithm

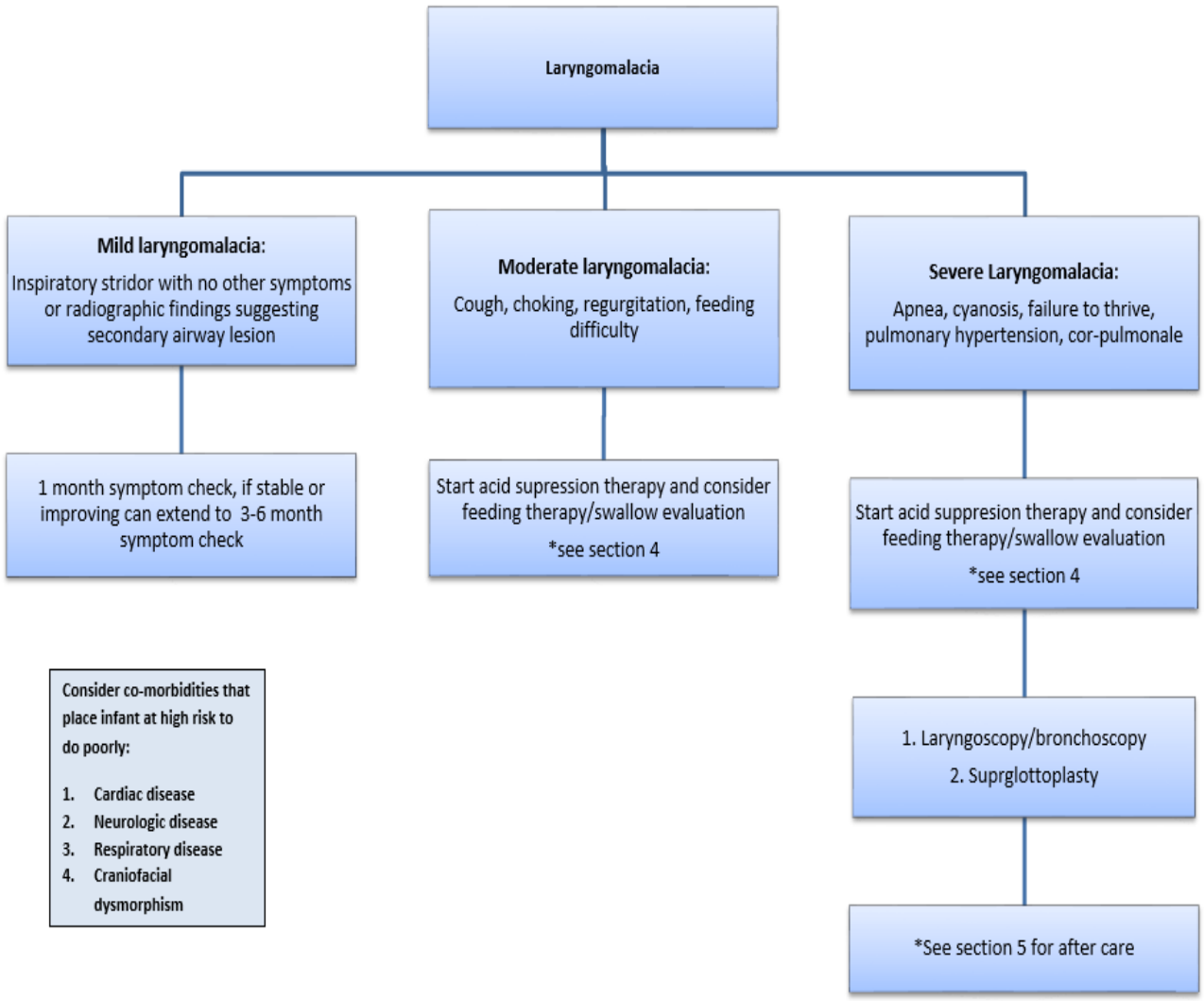
The initial presentation algorithm is designed to guide the initial evaluation of the infant presenting with inspiratory stridor. This may vary depending on what type of medical care setting the infant presents in. Flexible fibertoptic laryngoscopy (FFL) is important to confirm the diagnosis. Urgency of referral to an otolaryngologist is guided by severity of disease. Those with more severe disease may warrant expedited referral and those who have significant apnea/desaturations and/or inability to feed may warrant inpatient admission. Those infants who may be aspirating and/or have pulmonary disease may benefit from chest x-ray to further evaluate this. Those infants whose laryngoscopy findings are not commensurate with the severity of their symptoms may benefit from airway films to screen for a secondary airway lesion. Further recommendations are detailed in **Figure 1.**

Figure 1. Initial presentation algorithm



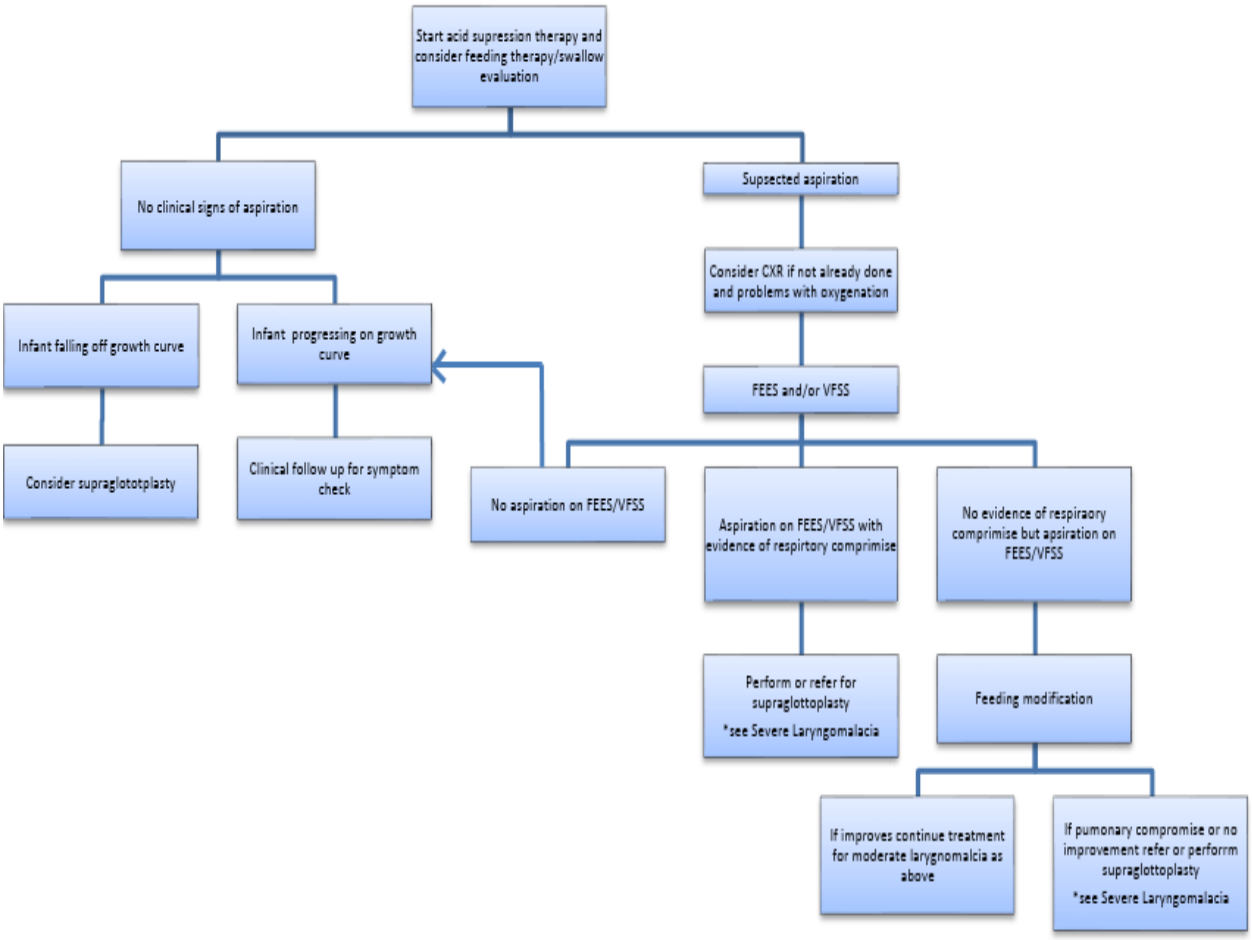
Section 3: Comprehensive care algorithm

The algorithm in **Figure 2** was designed to guide treatment for the infant that has been diagnosed with laryngomalacia, confirmed by FFL. This algorithm stratifies management decisions based on disease severity. The group suggests that the provider should recognize the presence of co-morbidities (**see figure 2**) that may lead to sub-optimal outcomes. Additionally, supraglottoplasty should be carefully considered in those with neurologic disease whose aspiration could be made worse by surgery. Variation in practice among the current group members remains, and the purpose of this section is to provide a list of reasonable options based on expert opinion.



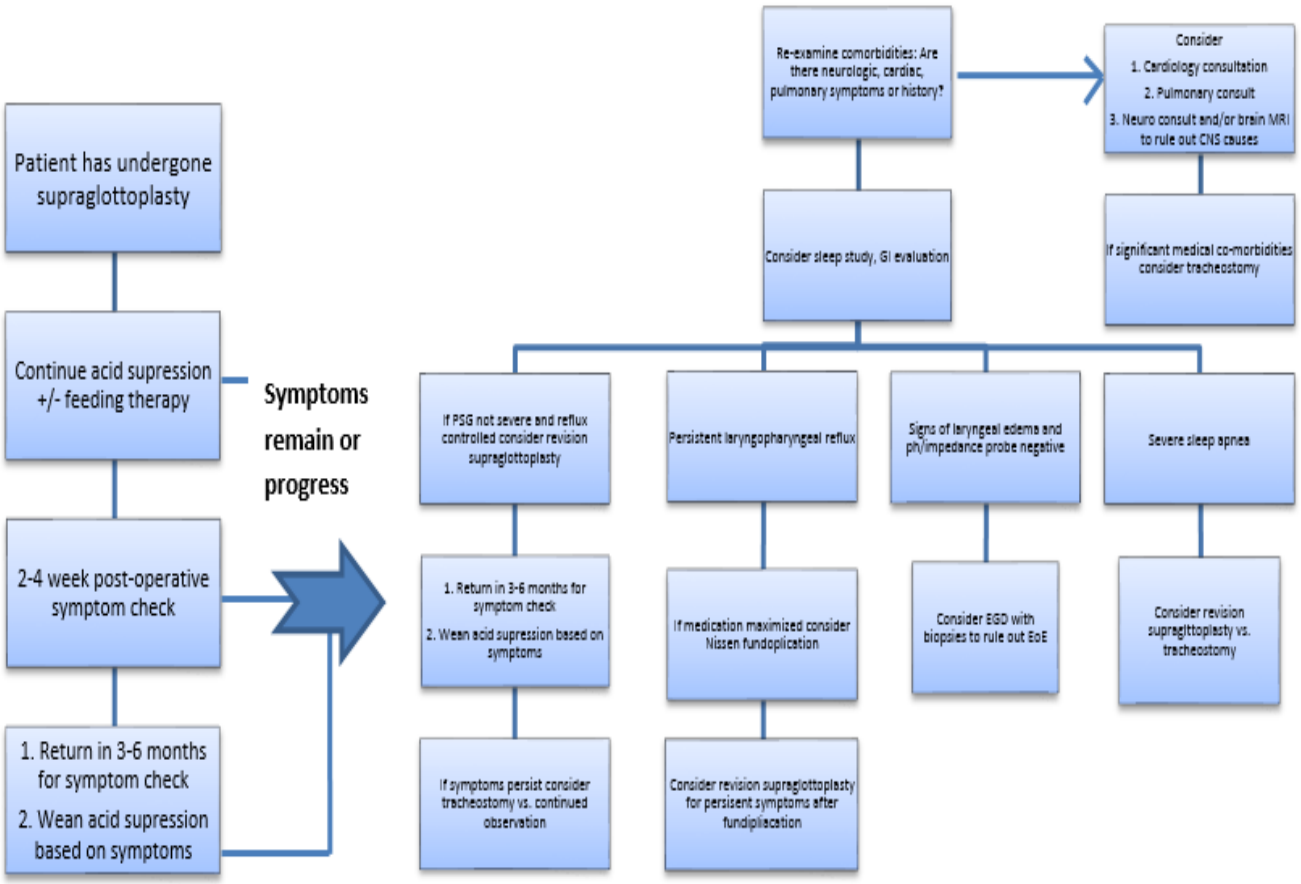
Section 4: Management of the difficult to feed infant with laryngomalacia

This algorithm in **Figure 3** outlines a guide for managing infants who have been diagnosed with laryngomalacia and are having difficulty with effectively and/or safely feeding. The algorithm may be best performed in conjunction with feeding or speech therapy services when available to guide feeding recommendations and therapy. Chest x-ray may be indicated in the infant if there are signs of respiratory compromise. Again, variation in practice among the current group members remains, and the purpose of this section is to provide a list of reasonable options based on expert opinion.



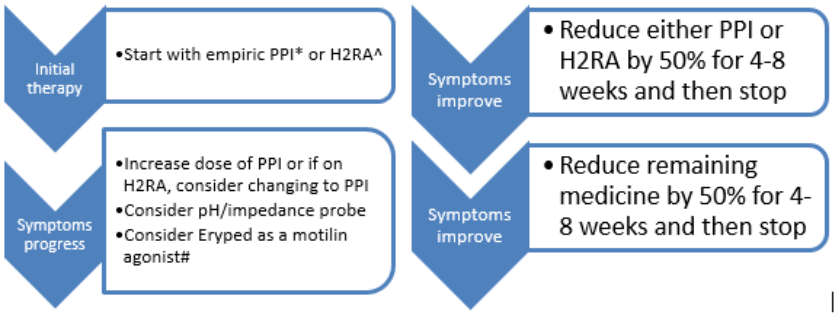
Section 5: Post-surgical treatment algorithm and persistent laryngomalacia

The algorithm displayed in **Figure 4** is intended to guide management in infants whom have undergone supraglottoplasty and is targeted towards those who have persistent laryngomalacia despite surgery. Time course for follow-up is variable amongst providers and is dictated by the severity of persistent symptoms in the patient. Persistent laryngopharyngeal reflux or undiagnosed eosinophilic esophagitis may drive persistent laryngeal edema leading to continued laryngomalacia. Confirming the presence or absence of obstructive sleep apnea may drive decision making towards surgical management vs. observation. Lastly, identifying and optimizing cardiac, neurologic, and pulmonary co-morbidities is paramount in managing the infant with persistent symptoms after supraglottoplasty, especially before considering revision surgery. Again, variation in practice among the current group members remains, and the purpose of this section is to provide a list of reasonable options based on expert opinion.



Section 6: Recommendations for acid suppression therapy

Figure 5 provides a "step-up" vs. a "step-down" regimen for managing acid suppression therapy. In a "step down" regimen, therapy can be started with both a proton pump inhibitor (PPI) and a histamine-2 blocker (H2RA) and then weaned to a single therapy if the patient improves. Conversely, the infant can be started conservatively on a single therapy and "stepped-up" to dual acid suppression if symptoms are not controlled. We suggest maintaining therapy for at least 3 months after initiation and a wean should not be considered until a diet can be safely tolerated from an aspiration standpoint. Consider gastroenterology evaluation and/or pH/impedance probe testing in those that are refractory to therapy. Additionally, erythromycin ethylsuccinate (Eryped) can be considered to improve gastrointestinal motility in refractory cases. There was great variation in practice among the current group members, and the purpose of this section is to provide reasonable options to guide the practitioner when using acid suppression in the setting of laryngomalacia.



* Preferred approach for children and adolescents, particularly when used empirically; QD dosing initially - AM dosing on empty stomach provides best acid suppression because H^+ pump less activated nocturnally, use PM dosing for nocturnal symptoms

^ Decrease acid production by 40-60% and well tolerated; use mandated before PPI trial by some insurance carriers; preferred for infants with non-life-threatening symptoms as H2RAs clinically better tolerated than PPIs

Low dose Erypred 200 (200 mg/5ml) or Erypred 400 (400mg/5ml) at 1-2 mg/kg/dose, 15 min before meals, up to 6 x per day as a motilin agonist to increase smooth muscle contraction

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