# ICD-10-CM Coding for Short Stature due to Endocrine Disorder:

## Prepared for ICD-10 Coordination and Maintenance Committee Meeting

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# Summary of Request

- Advanced research and technology necessitate a modification to the International Classification of Disease, Tenth Revision, Clinical Modification (ICD-10-CM) code for Short stature due to endocrine disorder (E34.3)
  - Specifically E34.3 should separately identify idiopathic short stature, constitutional short stature, primary IGF-I deficiency, and IGF-I (receptor) resistance
- This change is important for:
  - **Alignment with providers:** The Pediatric Endocrine Society and providers agree that updating this coding set would provide clarity and consistency
  - **Improve patient treatment coordination:** People diagnosed with short stature due to endocrine disorders receive different types of treatments depending on the cause of short stature and improved coding could help ensure they receive the correct treatment
  - **More accurate disease tracking:** Updating the codes would allow for more accurate tracking of the frequency of the disease and healthcare resource utilization



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# Overview of Short Stature

## Changes in normal growth patterns may be a sign of a pathologic condition

- Short stature is defined by a height that is at least 2 standard deviation scores (SDS) below the mean height of the age group

## Approximately 5% of children have short stature due to endocrine disorders

- Postnatally both GH and IGF-I drive statural growth through the pubertal growth spurt
- Short stature can develop when the GH/IGF-I axis is altered

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1. Barstow C, Rerucha C. Evaluation of Short and Tall Stature in Children. Am Fam Physician. 2015;92(1):43-50.



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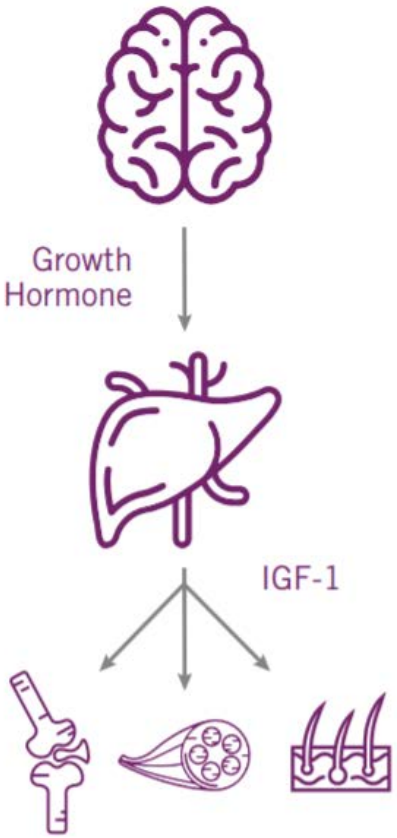
# Normal Growth Depends on the Growth Hormone and Insulin-like Growth Factor-I




Growth Hormone (GH) is released from the pituitary into circulation

GH interacts with its receptors on the liver (and other tissues) to stimulate release of IGF-I into circulation

IGF-I then interacts with its receptors in bone, skin, and muscle to stimulate growth

**When these pathways are altered, short stature can develop**

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GH: Growth Hormone; IGF-I: Insulin-like growth factor-1

1. Dehkhoda, Farhad, Christine MM Lee, Johan Medina, and Andrew J. Brooks. "The growth hormone receptor: mechanism of receptor activation, cell signaling, and physiological aspects." *Frontiers in Endocrinology* 9 (2018): 35.

IGF-1: Insulin-like growth factor; GH: Growth hormone



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# With A Broader Knowledge of Both Hormonal Interactions and Molecular Genetics, Short Stature Syndrome Has Become Better Defined

**Short stature due to endocrine disorder includes:**

- **Idiopathic Short Stature (ISS):** Short stature that is defined by height that is less than 2 SDS below the mean height for the corresponding population (e.g., age, gender)
- **Constitutional Short Stature:** Height below the 5th percentile, but normal height velocity. Height below the expected growth curve based on parental heights. Due to bone age delay growth continues beyond normal age of growth cessation and adult height ends up close to the target height range
- **Severe Primary IGF-I Deficiency:** Short stature defined by height and circulating IGF-I concentrations that are less than 3 SDS below the mean when compared to the corresponding population (e.g., age, gender) despite normal or elevated GH levels
- **IGF-I Resistance:** Short stature due to a defect in the type 1 IGF receptor and associated by variable degrees of intrauterine and postnatal growth retardation associated with normal or elevated serum IGF-I concentrations

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GH: Growth Hormone; ISS: Idiopathic Short Stature; IGF-1: Insulin-like growth factor-1 ; SDS: Standard deviation score

1. Insulin-like Growth Factor Deficiency. The Magic Foundation. Available at: www.magicfoundation.org/Growth-Disorders/Insulin-Like-Growth-Factor-Deficiency. Accessed July 2020; 2.Cohen P et al. *J Clin Endocrinol Metab* 2008;93(11):4210–4217. 2. Cohen P et al. *J Clin Endocrinol Metab* 2008;93(11):4210–4217. Available Here. Accessed August 2020. 3. Constitutional Growth Delay and Familial Short Stature: A Guide for Families. Pediatric Endocrine Society. Available Here. Accessed August 2020. 4. Grimberg A et al. *Clinical Practice Committee Publication* 2016;86(6):361-397. Available Here. Accessed August 2020. 5. Savage M et al. Frontiers in Endocrinology 2011; 2(95). Available Here. Accessed August 2020.



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# Differential Diagnosis Is Important for Patients with IGFD Patients

**Screening**

- Auxological parameters
- Circulating hormone concentrations

→ **Diagnosis**

- GH Deficiency (GHD)
- Idiopathic Short Stature (ISS)
- Constitutional Short Stature
- Primary IGF-I Deficiency (PIGFD)
- IGF-I Resistance

→ **Treatment**

- rhGH Replacement Therapy
- rhGH Supplementation Therapy
- Observation, androgen therapy, etc.
- rhIGF-1 Replacement Therapy
- No current FDA-approved therapy

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1. Grimberg A et al. *Horm Res Paediatr* 2016;86:361–397.



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# Updated Coding Is Crucial to Uniquely Identify Patients with Short Stature

## Rationale:

**Current Pediatric Endocrine Society Recommendations:**

- Separating out idiopathic short stature, constitutional short stature, primary IGF-I deficiency, and IGF-I resistance, would more closely align with the current Pediatric Endocrine Society recommendations for diagnosis and management of growth disorders

**More accurate disease tracking**:

- Updating the codes would allow for more accurate disease tracking to clarify the prevalence of the disorder as well as adequately support health resource utilization in the diagnosis

**Improve treatment**:

- People diagnosed with short stature due to endocrine disorders receive different types of treatments depending on the cause of short stature

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