

National Burden of Achondroplasia in Adults and Children: An Analysis of the National Inpatient Sample

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Er Chen ¹, Jessie T. Yan ², Eunice Chang ², Michael S. Broder ², Marian H. Tarbox ², Adelpha Abrahamson Larkin ¹, Wayne Pan ¹

OBJECTIVES	RESULTS	RESULTS (continued)	RESULTS (continued)
<ul style="list-style-type: none">Achondroplasia is the most common form of skeletal dysplasia and is associated with orthopedic, neurological, respiratory, and otolaryngologic complications.There are currently no approved pharmacological treatments for achondroplasia, and patients often require multiple surgeries.	<p>Demographic characteristics and hospital characteristics (Table 1)</p> <ul style="list-style-type: none">In 2017, there were 1,985 admissions of people with achondroplasia nationwide (1,090 adults and 905 children).Among adults, about one-third of the admissions were in the 31-49 year age range; among children, the majority of admissions were in the 0-4 year age group.	<p>Hospitalization LOS and cost (Figure 1)</p> <ul style="list-style-type: none">The average hospital LOS (95% CI) was 6.8 (5.7-8.0) days, with 7.1 (5.0-9.2) for children and 6.6 (5.5-7.7) for adults.Total mean costs (95% CI) per hospital admission were \$19,959 (\$16,801-\$23,118), with \$22,031 for children and \$18,899 for adults.	<p>NASS</p> <ul style="list-style-type: none">Compared to hospital admissions among children with achondroplasia where the 0-4 years age group accounted for the largest percentage of admissions, children ages 5-14 years accounted for the majority of outpatient surgical procedures (95.0% (95% CI) 93.0-97.0%).

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PRESENTED AT:



OBJECTIVES

- Achondroplasia is the most common form of skeletal dysplasia¹ and is associated with orthopedic, neurological, respiratory, and otolaryngologic complications²⁻⁵
- There are currently no approved pharmacological treatments for achondroplasia, and patients often require multiple surgeries throughout their lifetimes²⁻⁸
- There are no national estimates of the cost or burden of hospitalization and surgery in this population
- We aimed to use nationally representative data to estimate the cost and burden of hospital admissions for people with achondroplasia in the US
 - As a secondary objective, we also used nationally representative data to estimate the cost and burden of outpatient surgical procedures in this population

METHODS

Study design and data source

- Retrospective, cross-sectional cohort analysis of the National Inpatient Sample ([NIS] part of the Healthcare Cost Utilization Project [HCUP] sponsored by the Agency for Healthcare Research and Quality [AHRQ])
 - NIS is the largest all-payer inpatient healthcare database in the US, representing a 20% stratified sample of all discharges in the US and containing data from >7 million unweighted (>35 million weighted) hospital stays each year
- For secondary objective: The National Ambulatory Surgery Sample (NASS) is the largest all-payer national ambulatory/outpatient surgery database in the US, representing a 63% stratified sample of the US hospital-owned facilities performing ambulatory surgeries

Population

- Children (<18 years old) and adults (≥ 18 years old) hospitalized with a diagnosis of achondroplasia (ICD-10-CM code: Q77.4) from Jan to Dec in 2017

Study measures

- Patient and hospital characteristics
- Hospitalizations (number and type)
- Length of stay (LOS)
- Hospital cost

Statistical analysis

- Each observation (discharge) was weighted to represent national estimates
- Domain (subgroup) analysis was used to estimate variance due to the complex weighting of NIS and NASS
- Outcomes benchmarked against HCUP national mean values where possible
- As NIS only reports charges, the hospital-specific all-payer inpatient cost-to-charge ratio was used to estimate costs
- All data transformations and descriptive statistical analyses were performed using SAS© version 9.4

RESULTS

Demographic characteristics and hospital characteristics (Table 1)

- In 2017, there were 1,985 admissions of people with achondroplasia nationwide (1,080 adults and 905 children)
 - Among adults, about one-third of the admissions were in the 31-49 year age range; among children, the majority of the admissions were in the 0-4 year age range
- The majority of hospitals were urban and teaching hospitals, and were classified as large (per bed size category defined by HCUP)

Table 1. Demographic characteristics and primary payer for hospitalized patients with achondroplasia

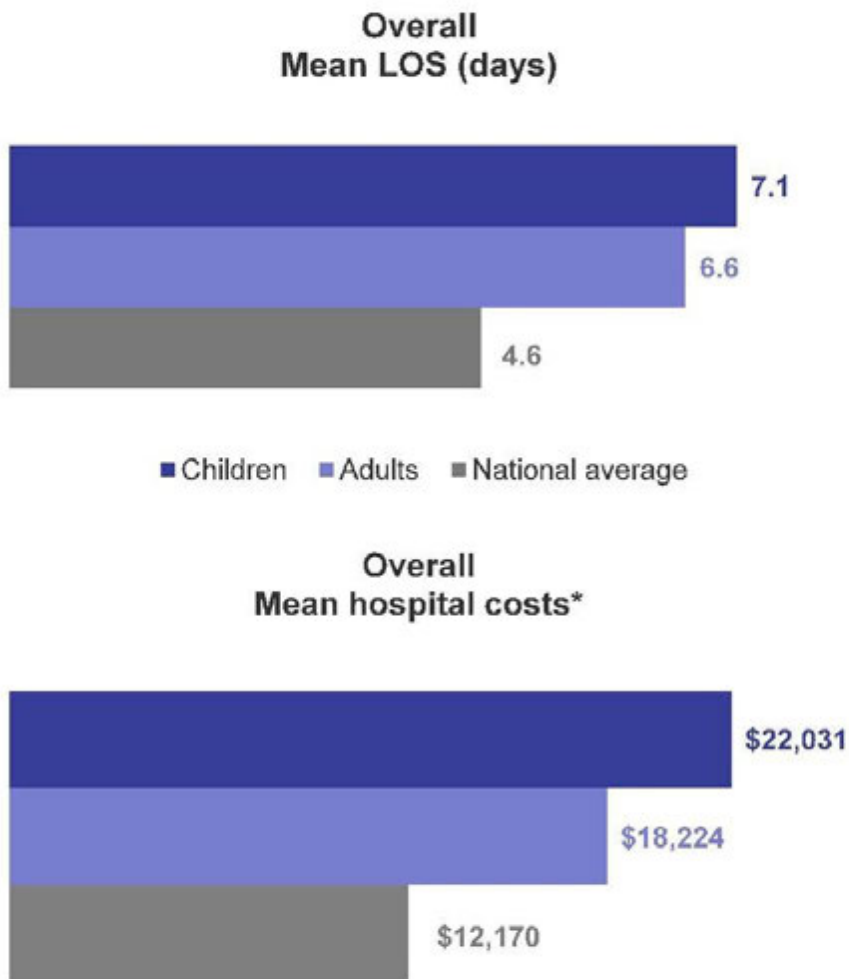
	Adults	Children
Weighted no. of admissions	1,080	905
Age, mean (95% CI)	45.9 (43.3 - 48.5)	4.0 (3.0 - 4.9)
Median (min: max)	43 (18: 87)	1 (0: 17)
Age group, % (95% CI)		
<1	n/a	48.1% (39.7% - 56.5%)
1-4	n/a	21.0% (15.0% - 27.0%)
5-14	n/a	22.7% (16.2% - 29.1%)
15-17	n/a	8.3% (3.5% - 13.1%)
18-30	25.9% (19.9% - 32.0%)	n/a
31-49	30.6% (24.4% - 36.7%)	n/a
50-64	25.5% (19.3% - 31.6%)	n/a
65+	18.1% (12.5% - 23.6%)	n/a
Female, % (95% CI)	62.5% (55.7% - 69.3%)	47.5% (39.5% - 55.5%)
Race, % (95% CI)		
White	55.6% (48.3% - 62.8%)	48.1% (39.6% - 56.5%)
Black	19.4% (13.8% - 25.1%)	13.3% (7.9% - 18.6%)
Hispanic	12.0% (6.9% - 17.1%)	16.0% (10.0% - 22.1%)
Other	6.5% (3.0% - 9.9%)	15.5% (9.3% - 21.6%)
Missing	6.5% (2.1% - 10.9%)	7.2% (2.7% - 11.7%)
Primary payer, % (95% CI)		
Medicare	45.4% (38.6% - 52.2%)	0.6% (0.0% - 1.6%)
Medicaid	24.5% (18.8% - 30.3%)	49.7% (41.6% - 57.8%)
Private (including HMO)	25.9% (20.0% - 31.9%)	43.6% (35.5% - 51.8%)
Self-pay	2.8% (0.6% - 5.0%)	1.7% (0.0% - 3.5%)
Missing/No charge/Other	1.4% (0.0% - 3.0%)	4.4% (1.5% - 7.4%)
Region of hospital, % (95% CI)		
Northeast	18.1% (13.0% - 23.1%)	13.8% (8.3% - 19.4%)
Midwest	30.6% (23.3% - 37.8%)	18.2% (11.4% - 25.1%)
South	31.5% (24.3% - 38.6%)	38.1% (29.3% - 46.9%)
West	19.9% (14.1% - 25.7%)	29.8% (20.6% - 39.0%)
Urban hospital, % (95% CI)	95.4% (92.3% - 98.4%)	97.2% (94.8% - 99.6%)
Teaching hospital, % (95% CI)	80.1% (74.6% - 85.6%)	88.4% (83.0% - 93.7%)
Bed size of hospital, % (95% CI)		
Small	19.4% (13.4% - 25.5%)	18.2% (11.2% - 25.3%)
Medium	19.0% (13.2% - 24.8%)	24.3% (16.5% - 32.1%)
Large	61.6% (54.3% - 68.9%)	57.5% (48.4% - 66.5%)

RESULTS (CONTINUED)

Hospitalization LOS and cost (Figure 1)

- The average hospital LOS (95% CI) was 6.8 (5.7-8.0) days, with 7.1 (5.0-9.2) for children and 6.6 (5.5-7.7) for adults
- Total mean costs (95% CI) per hospital admission were \$19,959 (\$16,801-\$23,118), with \$22,031 (\$16,311-\$27,752) for children and \$18,224 (\$15,157-\$21,292) for adults
- Patients with achondroplasia had a mean LOS that was 2.2 days longer and mean hospital costs that were \$7,789 greater than the national average

Figure 1. Mean length of stay and hospital costs* for patients admitted with achondroplasia compared to national average



LOS: length of stay.

* Physician costs were not included.

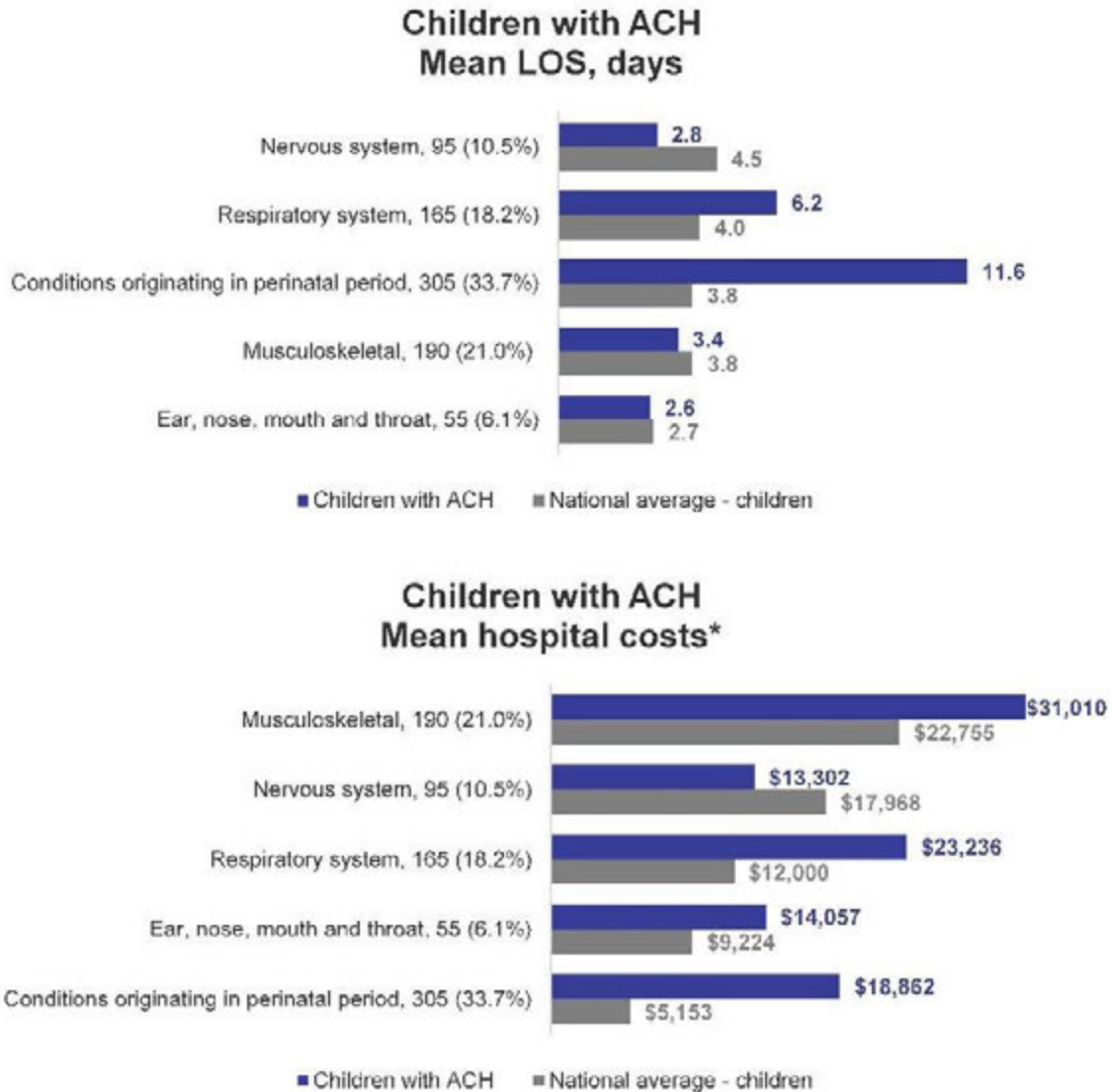
Note: National average based on HCUP values.

Major diagnostic category (Figures 2-3)

- The most frequent admission types were
 - neonatal care (33.7%) in children with mean LOS of 11.6 days, and
 - musculoskeletal (22.7%) in adults with mean LOS of 6.3 days (compared to national average of 3.9 days)

- Hospital costs for the majority of admissions among patients with achondroplasia were higher than national averages

Figure 2. Mean length of stay (days) and hospital costs* in children admitted with achondroplasia compared to national average† by major diagnostic category, n (%)

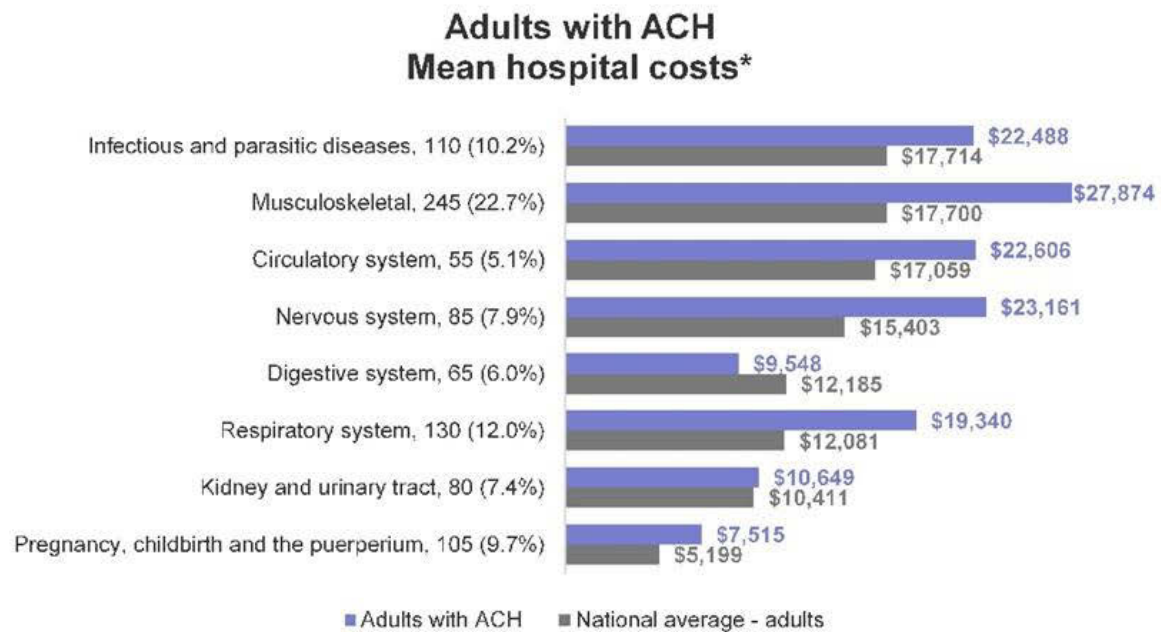
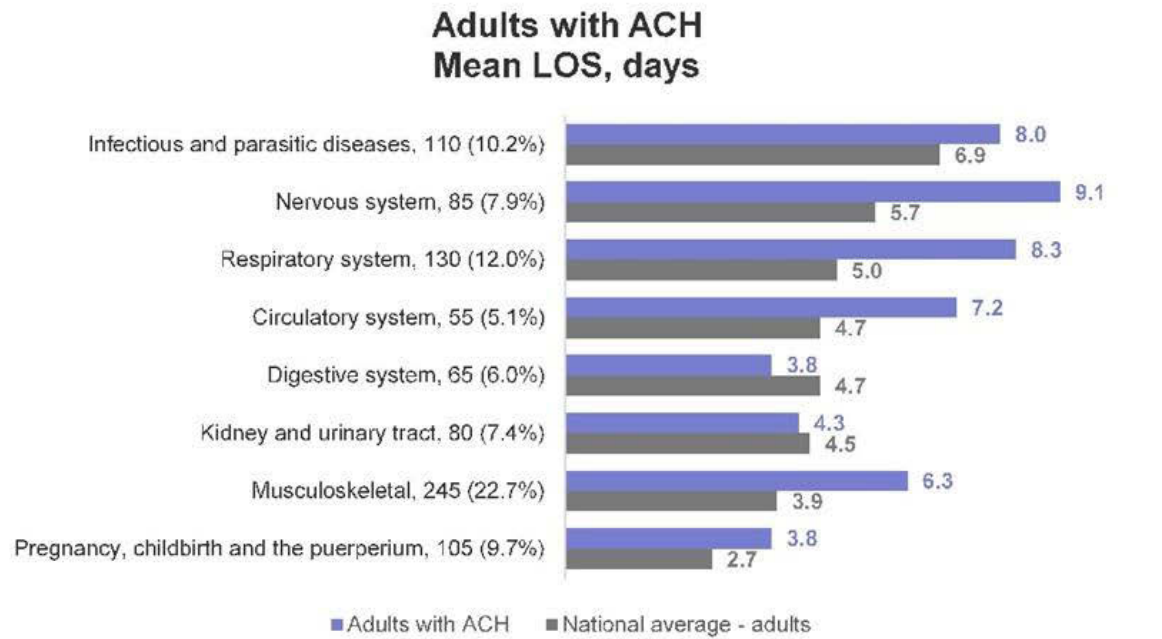


ACH: achondroplasia; LOS: length of stay.

* Physician costs were not included.

† National average based on Healthcare Cost and Utilization Project (HCUP) values.

Figure 3. Mean length of stay (days) and hospital costs* in adults admitted with achondroplasia compared to national average† by major diagnostic category, n (%)



ACH: achondroplasia; LOS: length of stay.

* Physician costs were not included.

† National average based on Healthcare Cost and Utilization Project (HCUP) values.

RESULTS (CONTINUED)

NASS

- Compared to hospital admissions among children with achondroplasia where the 0-4 years age group accounted for the largest percentage of admissions, children ages 5-14 years accounted for the majority of outpatient surgical procedures (56.9% [95% CI: 50.0%-63.9%]) (results not shown)
- The most common procedures were ear, nose, mouth and throat (ENT) (20.2% of all procedures for adults and 78.5% of all procedures for children) (**Figures 4A and 4B**)

Figure 4A. Ambulatory surgeries in children with a diagnosis of achondroplasia by major diagnostic category (%) – NASS

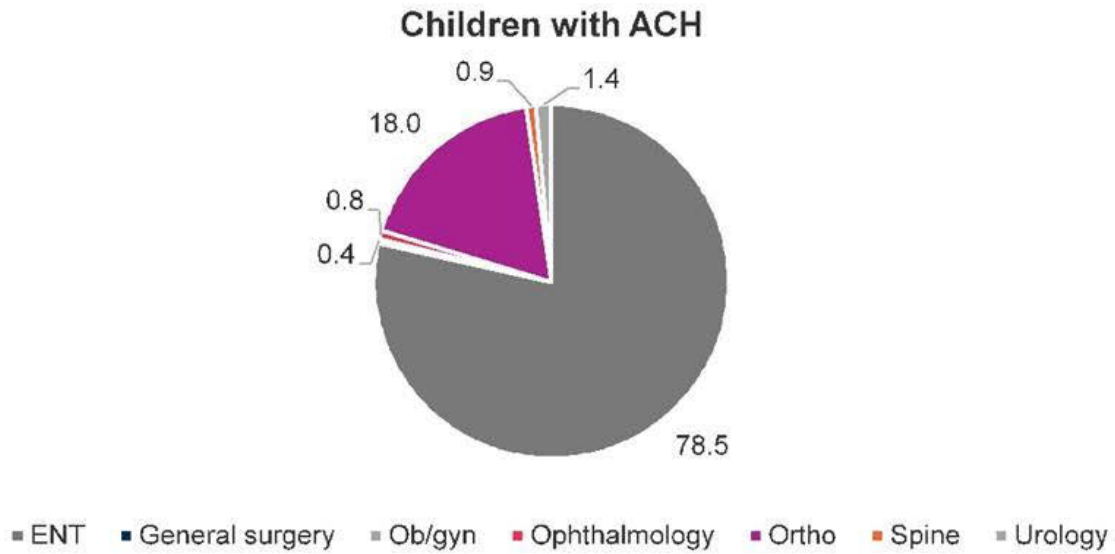
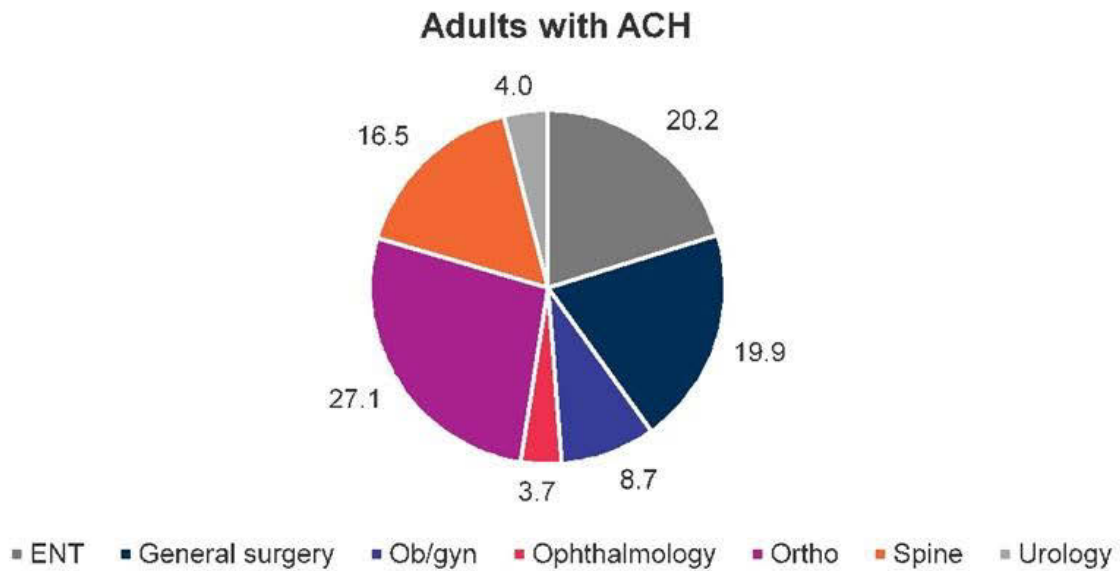


Figure 4B. Ambulatory surgeries with a diagnosis of achondroplasia by major diagnostic category (%) – NASS



ACH: achondroplasia; NASS: National Ambulatory Surgery Sample.

CONCLUSIONS

Limitations

- Patients with hypochondroplasia may be included, as the same ICD-10-CM diagnosis code is used for both conditions
- NIS data limited to hospital setting only; healthcare resource utilization and costs incurred outside of a hospital are not captured
- Although NIS and NASS are designed to represent discharges and major ambulatory surgery encounters nationally, rehabilitation and long-term acute care hospitals are excluded from NIS and non hospital-owned facilities from NASS
- Research is needed to further quantify total cost of care at patient level as well as impact of emerging new treatment on existing management of disease and costs

Conclusions

- Achondroplasia is associated with a wide range of complications frequently requiring hospitalization and surgical intervention
- In 2017 hospitalization cost alone was estimated to be \$40 million in the US
- Future work should be directed on impact of emerging disease modifying treatments on overall complications and costs

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