INTRODUCTION
Hutchinson-Gilford Progeria Syndrome (HGPS) is a rare, sporadic, autosomal dominant genetic disorder. The diagnosis is based on recognizing clinical features and the detection of a mutation on the LMNA gene. It is characterized by several clinical manifestations including short stature, craniofacial disproportion, premature graying of hair, alopecia, atrophied wrinkled skin, skeletal anomalies, and lipodystrophy. Accelerated, premature atherosclerosis leads to eventual fatal myocardial infarction, stroke, and congestive heart failure typically in the teenage years.

CASE PRESENTATION
We present a case of a 14 year old female with HGPS, HH, who presented with persistent angina and underwent a cardiac catheterization for progression of symptoms.

Patient reported a history of mixed dyslipidemia, hypertension, and moderate left ventricular hypertrophy with no previous TIA, stroke, or MI. EKG and ECHO were unremarkable with no evidence of prior MI and no wall motion abnormalities.

On physical exam, significant craniofacial disproportion with micrognathia was noted, with concern for securing the airway. Significant skeletal abnormalities affecting patient positioning also apparent.

Decision was made to proceed with monitored anesthesia care for the procedure.

After appropriate monitors were placed and supplemental oxygen provided by nasal cannula, propofol and ketamine were titrated while spontaneous ventilation was maintained.

During coronary angiography, the patient acutely developed bradycardia, hypotension, and ST depression. After successful resuscitation, further diagnostic coronary angiogram revealed a RCA with significant collateral perfusion of the left ventricle and no evidence of a left coronary artery.

Up for further diagnostic catheterization, no evidence of a left coronary artery origin could be found in the aorta.

DISCUSSION
Hutchinson-Gilford Progeria syndrome is a rare sporadic disorder with an incidence of approximately 1 in 8 million live births. Almost all individuals with HGPS are a result of a de novo autosomal dominant mutation. The cause is a mutation in LMNA gene and results in features characteristic of an accelerated aging process.

Several clinical manifestations of HGPS are important for appropriate preoperative evaluation of these patients.
• Growth: short stature and low weight; disproportionately large head.
• Airway: retrognathia, micrognathia, delayed eruption and loss of primary teeth, partial secondary tooth eruption, overcrowding of teeth.
• Body/skin: diminished subcutaneous fat; taut, dry skin, sclerodermatous skin especially over abdomen and upper thighs
• Skeletal/joints: pear shaped thorax, osteoarthritis, thin limbs, joint stiffness.
• CV: Severe progressive atherosclerosis manifested as angina, CHF, MI, stroke.

Anesthetic considerations may include:
• Careful titration of anesthetic agents.
• Preoperative plan for appropriate airway management.
• Positioning and padding of pressure points and joints.
• Evaluation of baseline ECG and ECHO.
• Carotid artery duplex scan.
• Avoid dehydration as stiffened vasculature may not tolerate hypotension.

HH was a HGPS patient with angina who was found to have single coronary artery perfusion. Upon occlusion of the single right coronary artery with the balloon during angiography, she experienced significant coronary ischemia with resulting bradycardia, hypotension, and ST depression. After successful resuscitation, further diagnostic coronary angiogram revealed a RCA with significant collateral perfusion of the left ventricle and no evidence of a left coronary artery.

Single coronary artery (SCA) is one of the most rare coronary anomalies with an incidence of 0.01 - 0.04% in the general population and 0.05 - 1.5% incidentally found in coronary angiograms. Congenital single coronary arteries were classified by Lipton in 1979 and typically have the left anterior descending and circumflex arteries arise from the RCA in different anatomic variations. This anomaly could be congenital but is likely the result of complete occlusion due to early atherosclerosis. No evidence of LAD or LCA could be found. We present this interesting case to alert providers of additional concerns during coronary catheterization in patients with HGPS.

REFERENCES

Table 1: Angiographic classification of a single coronary artery proposed by Lipton in 1979**

<table>
<thead>
<tr>
<th>Course of the transfer branch</th>
<th>Anatomical distribution</th>
<th>Origin location</th>
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<tr>
<td>A</td>
<td>Right sinus of Valsalva</td>
<td>L</td>
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<tr>
<td>B</td>
<td>Left sinus of Valsalva</td>
<td>L</td>
</tr>
<tr>
<td>C</td>
<td>Single coronary artery with normal right or left coronary artery (RC or LC)</td>
<td>L</td>
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<tr>
<td>D</td>
<td>After leaving the right or left sinus the single coronary artery crosses at the base of the heart to a large transverse trunk in order to supply the contralateral coronary artery</td>
<td>L</td>
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<tr>
<td>E</td>
<td>Single coronary artery arising from the right sinus, with the left anterior descending and circumflex arteries from separate coronary artery trunks instead of a single trunk immediately at the end of the right sinus</td>
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<tr>
<td>F</td>
<td>Anterior to the large vessels</td>
<td>R</td>
</tr>
<tr>
<td>G</td>
<td>Between the aorta and pulmonary artery</td>
<td>R</td>
</tr>
<tr>
<td>H</td>
<td>Posterior to the large vessels</td>
<td>R</td>
</tr>
<tr>
<td>I</td>
<td>Septal type (above the interventricular septum)</td>
<td>R</td>
</tr>
<tr>
<td>J</td>
<td>Combined type</td>
<td>R</td>
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