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Authors

Vadi, Marissa G
Ghazal, Elizabeth A
Malkin, Mathew R
[et al.](#)

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Continuous Non-Invasive Hemodynamic Monitoring in an Infant with Tetra-Amelia

1. Marissa G. Vadi, MD, MPH

- Title: Assistant Professor of Anesthesiology
- Affiliation: Department of Anesthesiology; Loma Linda University School of Medicine; Loma Linda, CA, U.S.A.
- Email: mvadi@llu.edu
- Conflicts of Interest: None

2. Elizabeth A. Ghazal, MD

- Title: Assistant Professor of Anesthesiology
- Affiliation: Department of Anesthesiology; Loma Linda University School of Medicine; Loma Linda, CA, U.S.A.
- Email: eghazal@llu.edu
- Conflicts of Interest: None

3. Mathew R. Malkin, MD

- Title: Assistant Professor of Anesthesiology
- Affiliation: Department of Anesthesiology; Loma Linda University School of Medicine; Loma Linda, CA, U.S.A.
- Email: mmalkin@llu.edu
- Conflicts of Interest: None

4. Abisola Ayodeji, MD

- Title: Pediatric Anesthesiology Fellow
- Affiliation: Department of Anesthesiology; Loma Linda University School of Medicine; Loma Linda, CA, U.S.A.
- Email: aayodeji@llu.edu
- Conflicts of Interest: None

5. Richard L. Applegate II, MD

- Title: Professor of Anesthesiology
- Affiliation: Department of Anesthesiology and Pain Medicine; University of California Davis, CA, U.S.A.
- Email: rapplegate@ucdavis.edu
- Conflicts of Interest: None

Name of Department(s) and Institution(s): Department of Anesthesiology, Loma Linda University School of Medicine, Loma Linda, CA, U.S.A. and Department of Anesthesiology and Pain Medicine; University of California Davis, CA, U.S.A.

Short Title: Hemodynamic Monitoring in Tetra-Amelia

Corresponding Author:

Marissa G. Vadi, MD, MPH

Department of Anesthesiology

Loma Linda University School of Medicine

11234 Anderson Street, Room 2532

Loma Linda, CA 92354

Phone: 909-558-4475

Fax: 909-558-4143

Email: mvadi@llu.edu

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4 **Abstract**
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8 Tetra-amelia syndrome is a congenital disorder associated with near or complete absence
9 of all four limbs. Non-invasive hemodynamic monitoring may be difficult or impossible in such
10 patients. We describe the use of a finger cuff blood pressure system for continuous non-invasive
11 blood pressure monitoring in an infant with near-complete tetra-amelia undergoing laparoscopic
12 gastrostomy tube placement. This case suggests the potential use of such a blood pressure
13 monitoring system for other patients with small extremities.
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24 **Number of words: 74**
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4 **Introduction**
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7 Tetra-amelia syndrome is a rare congenital disorder characterized by the absence of all
8
9 four limbs with other associated craniofacial, cardiopulmonary, neurologic, or urogenital
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11 anomalies. The absence of limbs may render non-invasive blood pressure (NIBP) measurement
12
13 with traditional blood pressure cuffs impossible. We report a case of continuous non-invasive
14
15 hemodynamic monitoring in an infant with tetra-amelia using a finger cuff blood pressure
16
17 measurement system (ClearSight™ System, Edwards, Irvine, CA, USA). The patient's mother
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19 reviewed the case report and provided written consent for publication.
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4 **Case Description**
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7 An 11-week-old, 4.2 kg infant with near complete tetra-amelia, sacral dysgenesis, and
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9 congenital micrognathia, hospitalized since birth at 39 weeks gestation in the Neonatal Intensive
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11 Care Unit (NICU) of our tertiary care hospital, was scheduled for laparoscopic gastrostomy tube
12
13 placement secondary to poor nipple and failure to thrive. Prior surgical procedures included
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15 mandibular distractor placement and insertion of a single lumen tunneled venous catheter at age
16
17 3 weeks, at which time fiberoptic intubation was performed with difficulty. The infant's bilateral
18
19 upper extremities and right lower extremity were completely absent, with only a rudimentary left
20
21 lower extremity appendage present (Figure 1). Hemodynamic monitoring was previously
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23 achieved via umbilical artery catheterization, which had been discontinued due to thrombotic
24
25 risk several weeks prior to the day of gastrostomy surgery. Subsequent attempts at NIBP
26
27 monitoring were unsuccessful, as standard neonatal blood pressure cuffs did not fit the patient's
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29 left lower extremity appendage. Pulse oximetry measurements were obtained from the distal
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31 appendage (Figure 1).
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39 A multidisciplinary discussion was held between pediatric anesthesiology, neonatology,
40
41 and pediatric surgery to determine the safest plan for intra- and post-operative hemodynamic
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43 monitoring. Full mandibular distraction had not yet been achieved, and there was a concern that
44
45 airway management attempts might be protracted. This concern heightened the desire to have
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47 reliable hemodynamic measurements. The patient was deemed a poor candidate for invasive
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49 arterial pressure monitoring after ultrasound examination revealed an absent right femoral artery,
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51 with underdeveloped left femoral and bilateral axillary arteries. Cerebral near-infrared
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53 spectroscopy (NIRS) was considered as a means to monitor trends in cerebral blood flow, but an
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4 appropriate sized sensor for this patient to allow use of this technology was not readily available
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7 in our operating room.
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10 The circumference of the patient's left lower extremity appendage was noted to be similar
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12 to that of an adult index finger. A size small finger cuff (ClearSight™ System, Edwards
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14 Lifesciences, Irvine, CA, USA), used in adults for non-invasive measurement of finger arterial
15
16 pressure, was placed on the left lower extremity appendage pre-operatively while the infant was
17
18 still in the NICU (Figure 2) and a reconstructed arterial pulse pressure waveform was obtained
19
20 (Figure 3). The patient, with the finger cuff blood pressure measurement system in place, was
21
22 then taken to the operating room for gastrostomy tube placement. After induction of general
23
24 anesthesia, spontaneous ventilation was maintained and a 3.5 uncuffed endotracheal tube was
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26 placed with moderate difficulty via video laryngoscope-assisted flexible fiberoptic intubation
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28 after 10 minutes of airway management. The anesthetic course was otherwise unremarkable.
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32 Continuous non-invasive blood pressure measurement was provided by the finger cuff system
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34 during the 45-minute surgical procedure and for 2 hours post-operatively. There was no
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evidence of vascular compromise or other injury to the left lower extremity appendage after cuff
discontinuation.

Discussion

Patients with severe limb abnormalities such as tetra-amelia present unique challenges to the anesthesiologist, as standard monitor placement is often problematic. Invasive blood pressure monitoring may be employed in such cases, but frequently target arteries are absent, aberrant, or underdeveloped. Superficial temporal artery cannulation via surgical cut-down under ketamine sedation has been described for hemodynamic monitoring during spinal fusion in a teenager with tetra-amelia.¹ However, this procedure would be difficult to accomplish in a young, uncooperative child without sedation or general anesthesia and thus was not attempted in our infant patient with a suspected difficult airway.

Few reports outline alternative strategies for non-invasive blood pressure monitoring in patients with significant limb deformities. Temporary penile blood pressure monitoring was used in a 3-year-old boy with severely shortened limbs unable to accommodate a standard blood pressure cuff.² Only intermittent cuff inflation was allowed and periodic penile inspection was required to rule out penile ischemia. After a 2-hour period, a femoral arterial catheter was placed with good correlation between the blood pressure cuff and arterial catheter. In the case we report, this would not have been a feasible option based on the patient's size.

General anesthetics have been performed in tetra-amelia patients with hemodynamic monitoring via carotid arterial Doppler³ and transesophageal Doppler.⁴ However, changes in Doppler sounds or pattern only allow for an approximation of blood flow trends, not adequacy of blood pressure, and the ability to detect subtle variations in sound intensity may vary by type of equipment and by operator. The use of NIRS to measure cerebral and somatic oxygenation

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4 trends in a patient with tera-amelia was described in a case report.¹ The authors hypothesized that
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6 a sustained drop in oxygenation is related to ischemic events that could be due to hypotension or
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8 blood loss. The primary use of NIRS in anesthetized infants and children has been to monitor
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10 cerebral oxygenation in children undergoing cardiac surgery but not as a sole monitor of
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12 perfusion adequacy.
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17 The system we employed uses a pneumatic finger cuff and infrared technology to detect
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19 pulsations that appear to correlate to blood pressure readings. This technology shows promising
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21 results in pediatric studies but has yet to be validated in the neonatal population.⁵ Although this
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23 technology may only monitor trends in patients such as ours, a provider may be able to use this
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25 to guide management in short cases where the use of traditional monitoring devices may not be
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27 feasible due to small extremity size. The minimum cuff size available and the need for adequate
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29 arterial pulsatility limit use in very small infants. Each cuff can only be used once, so cost of
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31 system usage increases if more than one cuff is required per anesthetic. Long term use should be
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33 avoided in small infants as there is an increased risk of limb edema and ischemia distal to the
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35 cuff position.⁵
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42 In conclusion, routine monitoring during anesthesia in patients with small or malformed
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44 extremities can pose a significant challenge to anesthesia providers. This case illustrates that use
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46 of a continuous noninvasive finger blood pressure system may allow successful blood pressure
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48 monitoring in infants with these conditions, and may be an alternative to continuous invasive
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50 blood pressure monitoring in short procedures without significant blood loss. Such systems
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58 ¹ Anesthetic Management of a Patient with Tetra-Amelia Syndrome. Available at:
59 https://www.webmedcentral.com/wmcpdf/Article_WMC00592.pdf. Accessed 3/7/16.
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could potentially replace arterial cannulation in children who do not have shock or hemorrhage conditions. Further studies are required to validate this technology in the pediatric population.

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4 **Figure Legends**
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8 **Figure 1: Frontal View of Patient**
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10 The patient was born with complete absence of the bilateral upper extremities and right lower
11 extremity. Only a rudimentary left lower extremity appendage was present. Mandibular
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13 distractors were previously placed for correction of severe micrognathia and remained *in situ* at
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15 the time of presentation for laparoscopic gastrostomy tube placement.
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24 **Figure 2: Finger Cuff Positioning**
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26 A size small finger cuff (ClearSight™ System, Edwards Lifesciences, Irvine, CA, USA) was
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28 placed on the distal portion of the patient’s left lower extremity appendage. Pulse oximetry
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30 measurements were obtained from the proximal portion of the left lower extremity appendage.
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37 **Figure 3: Arterial Pulse Pressure Waveform**
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39 The reconstructed arterial pulse pressure waveform obtained from the left lower extremity
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41 appendage finger cuff is displayed above. This waveform was monitored throughout the
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43 intraoperative and postoperative period.
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