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Rare Triad of Atrioventricular Septal Defect, Single Atrium, and Dextrocardia Presenting with Acute Decompensation and Recurrent Supraventricular Tachycardia

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Abstract

Background and Purpose: Atrioventricular canal defects (AVSD) represent 3-7% of congenital heart malformations (incidence 0.24-0.31/1,000 live births). When combined with dextrocardia and single atrium, this triad becomes exceptionally rare with only scattered case reports globally. Unoperated complex congenital heart disease typically progresses toward pulmonary vascular disease and poor outcomes. Current guidelines recommend surgical repair at 3-6 months of age to prevent irreversible complications. This case documents an acute presentation of this rare triad in an adolescent with unusual natural history.

Case Presentation: A 12-year-old female with AVSD, single atrium, and dextrocardia presented with 3 days of progressive respiratory distress, fever (38.5-39.2°C), cough, and vomiting. Baseline oxygen saturation was 88% on room air. On presentation: heart rate 146 bpm, respiratory rate 38 breaths/min, blood pressure 106/68 mmHg, oxygen saturation 88%. Examination revealed respiratory distress with retractions, hepatomegaly (5 cm below right costal margin), and altered mental status. Cardiac monitoring documented supraventricular tachycardia at 212 bpm. Laboratory studies: WBC 13,200/µL, hemoglobin 15.8 g/dL, Troponin I 0.08 ng/mL, BNP 485 pg/mL. Arterial blood gas: pH 7.28 (acidemia), pCO2 48 mmHg (hypercapnia), pO2 62 mmHg (hypoxemia). Chest radiography showed cardiomegaly, dextrocardia, and bilateral pulmonary edema. Echocardiography confirmed primum ASD (28 mm), VSD (18 mm), single atrium, dextrocardia, and pulmonary artery pressure 58 mmHg.

Management and Outcome: Intravenous adenosine (0.1 mg/kg = 3 mg) terminated SVT within 8 seconds. Supplemental oxygen (FiO2 40%) improved saturation to 92%. Furosemide (32 mg IV) and spironolactone (32 mg IV) initiated for diuresis. Ceftriaxone (80 mg/kg/day = 1,280 mg IV Q12H) and oseltamivir (60 mg BID) prescribed. Over 5-day hospitalization: dyspnea resolved, hepatomegaly regressed, oxygen saturation improved to 94%, ABG normalized, no SVT recurrence. Discharged on oral furosemide (32 mg BID), spironolactone (32 mg OD), acetaminophen (480 mg Q6H PRN), and home oxygen (1-2 L/min).

Conclusion: Unoperated complex congenital heart disease can compensate into adolescence but remains vulnerable to acute decompensation with respiratory infection and arrhythmia. Standard acute management effectively reversed decompensation, yet definitive surgical repair is essential to prevent irreversible pulmonary vascular disease and improve long-term survival.

Keywords: Atrioventricular septal defect; Congenital heart disease; Dextrocardia; Supraventricular tachycardia; Pulmonary hypertension

Introduction

Atrioventricular septal defects (AVSD), also known as atrioventricular canal defects, represent a well-characterized spectrum of congenital cardiac malformations resulting from incomplete development of the endocardial cushions. [1] AVSD accounts for approximately 3-7 percent of all congenital heart defects, with an incidence of 0.24-0.31 per 1,000 live births. [1] Complete AVSD is characterized by both atrial and ventricular septal defects with a common atrioventricular valve, resulting in significant left-to-right shunting at both atrial and ventricular levels. [1] This produces increased pulmonary blood flow and right heart volume overload, leading to pulmonary vascular disease development. [1,9]

The natural history of untreated complete AVSD is well-established: approximately 50 percent of patients die during infancy from congestive heart failure or infection, while survivors develop irreversible pulmonary vascular obstructive disease (PVOD) within two decades. [1,9] Current surgical guidelines strongly recommend early definitive repair between 3-6 months of age. [9,15] Modern outcomes demonstrate operative mortality of 2.6-2.9 percent with excellent long-term functional results. [14]

However, this case involves a 12-year-old with an exceptionally rare combination: complete AVSD plus dextrocardia (1 in 12,000-28,571 pregnancies)[2,3] plus single atrium (complete absence of interatrial septation). This triad has been reported in only a handful of cases worldwide.[1,2,79] The patient has remained surgically uncorrected throughout her 12-year lifespan—a remarkable deviation from standard management that reflects both unusual disease heterogeneity and access-to-care barriers.[15,16] At age 12, she presented with acute respiratory distress complicated by recurrent supraventricular tachycardia (SVT), her second episode in one year.[5,6,7,8] Single atrium creates a particularly arrhythmogenic substrate by eliminating normal conduction pathways.[5,17]

This case is clinically significant for multiple reasons. First, it documents an extraordinarily rare anatomic triad with limited literature representation.[1,2,3,7,9] Second, it illustrates natural history of unoperated complex disease extending into adolescence, suggesting disease heterogeneity not captured in existing surgical outcome studies.[1,9,11] Third, it demonstrates how respiratory infection precipitates acute hemodynamic collapse in compensated complex disease.[10,11,12,13] Fourth, it documents successful acute management with adenosine and diuretics in a highly unusual anatomic setting.[6,7,8] Finally, it emphasizes the critical temporal window for surgical intervention before irreversible PVOD develops. [9,11,18] At age 12 with already-present moderate pulmonary hypertension, this patient represents an urgent intervention candidate.[9,18]

Patient Information

Demographics: 12-year-old female, Indian ethnicity, rural Maharashtra, lower-middle-class background with limited access to specialized cardiac care.

Past Medical History: Complex congenital heart disease (AVSD + single atrium + dextrocardia) diagnosed at 6 months of age via echocardiography. Family declined early surgical repair due to financial constraints and limited understanding of natural history. One year prior (November 2023): hospitalization for acute respiratory distress with fever; SVT diagnosed (rate 198 bpm) and terminated with IV adenosine; discharged without antiarrhythmic prophylaxis. No chronic cardiac medications. Recurrent upper respiratory tract infections (3-4 times annually). Baseline oxygen saturation 85-88%. Exercise intolerant; dyspnea on minimal exertion. Feeding difficulties in infancy requiring 18 months nutritional support.

Family History: Mother healthy; father with controlled hypertension. Siblings (ages 15, 13) healthy with no cardiac or genetic conditions. Parents not consanguineous. No family history of CHD, genetic syndromes, or sudden death.

Psychosocial: Irregular school attendance due to health issues. Unable to participate in physical education or competitive sports. Quality of life significantly impaired by dyspnea and activity limitations. Parents anxious about prognosis; previously hesitant regarding surgery but now more receptive after acute presentation.

Case Presentation

Chief Complaint: "Progressive difficulty breathing for 3 days with fever and cough."

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Illness Timeline:

Date	Day	Key Events
Nov 12	Day 0	Evening: Fever 37.8°C, dry cough, rhinorrhea, lethargy onset
Nov 13	Day 1	Fever 38.5°C, dyspnea on exertion (stairs), first vomiting episode
Nov 14	Day 2	Fever 39.2°C, dyspnea at rest, marked accessory muscle use, 4-5 vomiting episodes
Nov 15	Day 3 AM	Continuous dyspnea at rest, severe accessory muscle use, marked lethargy
Nov 15	Day 3 PM	2:15 PM: Presented to ED; 4:30 PM: SVT episode (212 bpm), adenosine given

Respiratory Symptoms: Progressive dyspnea from exertional (Day 1) to resting (Day 3). Marked use of subcostal and intercostal accessory muscles. Respiratory rate 35-40/min. Bilateral decreased chest expansion, more on left. Persistent dry cough every 15-20 minutes. No hemoptysis. No wheezing.

Associated Symptoms: Moderate-grade intermittent fever (37.8-39.2°C), worse afternoons/evenings. Post-tussive non-bilious, non-projectile vomiting increasing from 1-2 episodes (Days 1-2) to 4-5 daily (Day 3). Generalized weakness; bedbound. Progressive lethargy: subtle Day 1 → markedly pronounced Day 3 (responds only to loud voice).

Negative History: No cyanotic episodes, palpitations, chest pain, syncope, presyncope, trauma, recent travel, medication ingestion, or seizures.

Clinical Findings

Table 2: Vital Signs On Presentation

Parameter	Value	Normal Range	Interpretation
Temperature	38.9°C	36.5-37.5°C	Moderate fever
Heart Rate	146 bpm (212 SVT)	60-100 bpm	Tachycardia
Respiratory Rate	38/min	18-25/min	Tachypnea
Blood Pressure	106/68 mmHg	95-105/55-70 mmHg	Borderline elevated
O2 Saturation	88% (RA)	≥95%	Hypoxemia
Capillary Refill	2.5 sec	<2 sec	Mildly delayed

Cardiovascular Exam: Apex beat right 5th ICS (dextrocardia confirmed). Hyperdynamic precordium. Normal S1; physiologically split S2. Grade 3/6 pansystolic murmur at left lower sternal border (VSD). Bounding peripheral pulses. Markedly elevated JVP (visible at jaw angle supine). No peripheral edema.

Respiratory Exam: Visible subcostal/intercostal retractions. Bilateral decreased chest expansion (left > right). Bilateral fine end-inspiratory crackles (bases prominent) consistent with pulmonary edema. Diminished air entry on left. No wheezing or rhonchi.

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Productive thin, non-purulent, non-blood-tinged sputum.

Abdominal Exam: Mild distension. Hepatomegaly: liver edge 5 cm below right costal margin (normal <2 cm). Smooth, firm liver; mild tenderness. No splenomegaly. Soft abdomen; mild supraumbilical tenderness; no peritoneal signs. Normal bowel sounds.

Neurological Exam: Lethargy with slow responses. All cranial nerves II-XII intact. Pupils equal, round, reactive. Motor 5/5 all extremities. Sensory intact. Reflexes 2+ symmetric. No meningeal signs. Altered sensorium attributed to: hypoxemia (SpO2 88%), hypercapnia (pCO2 48), reduced cerebral perfusion from SVT (212 bpm), metabolic acidosis (pH 7.28), and systemic illness.

Diagnostic Assessment

ECG: Sinus rhythm 118-126 bpm with left axis deviation (characteristic of AVSD). PR interval 0.16 sec (normal). QRS 0.10 sec (upper normal). QT 0.42 sec (normal). Right atrial enlargement (P waves ≥2.5 mm in II). RV hypertrophy (R wave V1 >7 mm; R:S ratio >1; rsR' pattern). LV hypertrophy (SV1+RV5=35 mm). ST downsloping in lateral leads; T wave inversions in II, III, aVF (RV strain pattern). SVT episode: 212 bpm, narrow QRS (0.08 sec), P waves buried in T wave. Terminated with IV adenosine.

Chest Radiography: Cardiomegaly (cardiac/thoracic ratio 65%, normal <50%). Dextrocardia confirmed. Boot-shaped cardiac configuration with RV prominence. Pulmonary plethora with increased vascular markings. Bilateral symmetric pulmonary edema: "butterfly" perihilar pattern, air bronchograms, Kerley B lines peripherally. Blunted costophrenic angles (small bilateral effusions). Lower lobe predominant interstitial infiltrates. No focal pneumonic consolidation, pneumothorax, or pneumomediastinum.

Echocardiography:

Table 3: Echocardiographic Measurements

Parameter	Measurement	Normal Range	Interpretation
Situs	Solitus	Normal	Normal visceral arrangement
Cardiac Position	Dextrocardia	Right side	Confirmed
Atrial Septation	Completely absent	Septum present	SINGLE ATRIUM (cor biloculare)
RA Dimension	55 mm	<40 mm	Severely dilated (138% normal)
LA Dimension	42 mm	28-35 mm	Dilated
RV EDD	42 mm	25-30 mm	Dilated (147% normal)
LV EDD	48 mm	40-47 mm	Upper normal limit
LV EF	52%	≥60%	Reduced
Primum ASD	28 mm	<15 mm	Large
VSD	18 mm	<15 mm	Moderate-to-large
Common AV Valve	Present	Separate MV+TV	Abnormal
TR	2+/4+	Trace	Moderate regurgitation
MR	1+/4+	Trace	Mild regurgitation
PA Systolic Pressure	58 mmHg	<25 mmHg	Moderate pulmonary hypertension

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Shunt assessment: Large left-to-right shunt across ASD (28 mm) and VSD (18 mm); bidirectional flow throughout cardiac cycle. Common AtrioVentricular (AV) valve with moderate TR and mild MR. Main PA dilated (22 mm; normal 16-18). RV function grossly preserved. No pericardial effusion or thrombus.

Laboratory Findings:

Table 4: Complete Blood Count

Parameter	Result	Normal Range
WBC	13,200/µL	4,500-11,000
Neutrophils	78%	50-70%
Hemoglobin	15.8 g/dL	11.5-15.5
Hematocrit	48%	34-45%
Platelets	285,000/µL	150,000-400,000

Elevated WBC with left shift (acute infection). Polycythemia (Hb 15.8, Hct 48%) from chronic cyanosis. Normal platelets (no DIC).

Table 5: Serum Chemistry

Parameter	Result	Normal Range
Sodium	135 mEq/L	135-145
Potassium	4.2 mEq/L	3.5-5.0
Chloride	102 mEq/L	96-106
BUN	24 mg/dL	7-20
Creatinine	0.9 mg/dL	0.7-1.3
Glucose	118 mg/dL	70-100

All electrolytes normal. Mildly elevated BUN (dehydration/reduced renal perfusion). Creatinine normal (renal function preserved). Stress hyperglycemia.

Table 6: Liver Function Tests

Parameter	Result	Normal Range
ALT	68 U/L	7-56
AST	82 U/L	10-40
Total Bilirubin	1.2 mg/dL	<1.2
Direct Bilirubin	0.4 mg/dL	<0.3

Mildly elevated ALT/AST from hepatic congestion and ischemia. Borderline bilirubin reflecting hepatic stress.

Table 7: Cardiac Biomarkers

Parameter	Result	Normal Range
Troponin I	0.08 ng/mL	<0.04
BNP	485 pg/mL	<100

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Elevated troponin reflecting myocardial stress from demand ischemia. Markedly elevated BNP indicating significant heart failure.

Table 8: Arterial Blood Gas (Room Air)

Parameter	Result	Normal Range
pH	7.28	7.35-7.45
pCO2	48 mmHg	35-45
pO2	62 mmHg	80-100
HCO3-	22 mEq/L	22-26
Lactate	2.8 mmol/L	0.5-1.5

Acidemia (combined respiratory + metabolic). Hypercapnia from ventilatory inadequacy. Hypoxemia from pulmonary edema. Elevated lactate indicating tissue hypoperfusion.

Microbiology: Blood cultures: No growth (rules out sepsis/endocarditis). Respiratory viral testing not performed; clinical presentation strongly suggestive of viral respiratory infection (RSV or rhinovirus).

Therapeutic Interventions And Hospital Course

Emergency Department Management (Day 1):

SVT (HR 212 bpm) documented on continuous cardiac monitoring. IV access established; labs drawn (CBC, CMP, ABG, troponin, BNP, blood cultures).

Medication Administration - Corrected Dosing: - **Adenosine:** 0.1 mg/kg = **3 mg** IV rapid bolus + 5 mL saline flush → SVT terminated in 8 seconds (HR decreased to 118 bpm sinus rhythm) - **Furosemide:** 1 mg/kg = **32 mg** IV bolus → urine output improved - **Spironolactone:** 1 mg/kg = **32 mg** IV → potassium-sparing - **Oxygen:** Nasal cannula 3 L/min → FiO2 40% → SpO2 improved to 92% - **Ceftriaxone:** 80 mg/kg/day = **1,280 mg IV Q12H** → empiric coverage - **Oseltamivir: 60 mg PO BID** × 5 days → viral infection suspected (appropriate for <40 kg) - **Acetaminophen: 480 mg Q6H PRN** (15 mg/kg × 32 kg) → fever management

Head of bed elevated 30-45°. Transferred to pediatric high-dependency unit.

Hospital Course Days 2-5:

TABLE 9: DAILY PROGRESSION

Day	Date	HR	RR	Temp	SpO2	O2	Status	Events
1	Nov 15	146→ 118	38	38.9	88→92	40%	Acute	SVT, adenosine given
2	Nov 16	110-130	32	38.2	93	2L NC	Improving	Urine 2.8 mL/kg/hr
3	Nov 17	118-126	28-30	37.5	94	1.5L NC	Improved	ABG normalized
4	Nov 18	102-115	26	36.8	94-95	1L NC	Visibly better	PO furosemide started
5	Nov 19	105-112	25	37.0	89-91	Home O2	Discharge ready	Weight loss 1.7 kg (5.3%)

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Day 2-3: No SVT recurrence. Respiratory status improved (RR 32→28, SpO2 93→94%). Fever decreased (38.2→37.5°C). Hepatomegaly regressed (5 cm→3 cm). Mental status improved (more alert, interactive). Repeat labs (Day 3): Troponin 0.04 (improved); BNP 285 (improved); ABG normalized (pH 7.36, pCO2 42, pO2 76).

Day 4-5: Vital signs stable. Furosemide transitioned to oral 32 mg BID. Spironolactone continued 32 mg OD. Chest X-ray showed marked pulmonary edema improvement; cardiomegaly persisted.

TABLE 10: DISCHARGE MEDICATIONS & INSTRUCTIONS (CORRECTED)

Medication	Dose	Frequency	Indication	Notes
Furosemide	**32 mg**	Twice daily	CHF management	1 mg/kg BID; 64 mg/day total
Spironolactone	**32 mg**	Once daily	Aldosterone antagonism	1 mg/kg
Acetaminophen	**480 mg**	Q6H PRN	Fever/pain control	15 mg/kg/dose; max 1,920 mg/day
Oxygen Therapy	**1-2 L/min**	Continuous at home	Maintain SpO2	Via nasal cannula; goal SpO2 92-95%

Discharge Instructions:

Fluid/Sodium Restriction: 1,000-1,200 mL/day fluid; <1 gram sodium/day.

Monitoring: Daily weights (same time each morning); report gain >0.5 kg/day. Monitor SpO2 regularly.

Activity: Light only; no strenuous exercise or sports

Discussion

Opening Summary: This case documents a 12-year-old with an exceptionally rare cardiac anatomy triad (AVSD + dextrocardia + single atrium) who presented with acute hemodynamic decompensation triggered by respiratory infection and complicated by recurrent SVT.[1,2,3] Despite profoundly complex unoperated heart disease, she achieved surprising baseline hemodynamic compensation through unknown adaptive mechanisms.[9] However, acute illness rapidly unmasked her limited cardiac reserve, necessitating intensive intervention. Standard acute management (adenosine, diuretics, oxygen, antibiotics) successfully reversed decompensation, but the fragility of this compensation emphasizes the critical urgency for definitive surgical intervention.[9,11,15,18]

Key Finding 1: Exceptional Rarity of Cardiac Anatomy Triad

This combination of three major cardiac anomalies has been reported in only a handful of cases globally.[1,2,3] AVSD occurs in 0.24-0.31/1,000 births (3-7% of CHD).[1] Dextrocardia occurs in 1 in 12,000-28,571 pregnancies.[2,3] Single atrium is extraordinarily rare.The convergence of all three anomalies is exceptionally uncommon and merits documentation for clinician education and recognition.[1,2,3] Comprehensive echocardiographic characterization with detailed measurements provides valuable reference data for rare case management.[1,9,18]

Key Finding 2: Unusual Natural History of Unoperated Complex Disease

Survival to age 12 with unoperated complete AVSD defies typical natural history where 50% die in infancy and most remaining patients develop PVOD by adolescence.[1,9] Eisenmenger syndrome demonstrates: 5-year survival 73%, 10-year survival 50%, mean age at death 17.4 years.[11] This patient's baseline stability suggests either: (1) subclinical disease not yet symptomatic; (2) protective physiologic mechanisms (optimized polycythemia, increased cardiac output); (3) anatomic factors of

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single atrium protecting against PVOD progression; or (4) inadequate documentation of subclinical dysfunction.[9,11,18] This heterogeneity deserves investigation.[1,9,11]

Key Finding 3: Respiratory Infection as Acute Decompensation Precipitant

Children with significant unoperated CHD demonstrate 3-4 fold increased susceptibility to severe respiratory infection complications compared to normal children.[10,11,12,13] Mechanistically: baseline pulmonary edema narrows margin between resting capacity and maximal effort;[9,14] respiratory viral infection causes mucosal edema, mucus production, increased airway resistance;[10,11,12,13] fever increases metabolic oxygen demand when delivery is already compromised;[9] infections trigger arrhythmias (this patient experienced SVT).[5,6,7,8] The convergence of chronic pulmonary overcirculation, acute viral infection, metabolic stress, and SVT created perfect cascade toward crisis.[9,10,11,12,13] This case illustrates why respiratory infections in unoperated CHD warrant high clinical suspicion for serious deterioration.[10,11,12,13]

Key Finding 4: Recurrent SVT and Abnormal Atrial Anatomy

Single atrium creates arrhythmogenic substrate by eliminating normal atrial septation and conduction pathways, predisposing to reentrant circuits.[5,17] Recurrence within one year strongly suggests persistent underlying arrhythmia mechanism unlikely to spontaneously resolve.[5,6,7,8] SVT first-line therapy (adenosine) achieved 72-79% success rate in pediatric populations.[6,7,8] This patient's adenosine response (termination in 8 seconds) was excellent.[6,7,8] However, recurrence pattern necessitates: (1) electrophysiologic study to characterize mechanism; (2) consideration of catheter ablation (84% success in pediatric patients); (3) prophylactic antiarrhythmic therapy (digoxin, propranolol, amiodarone); or (4) most importantly, definitive surgical repair normalizing anatomy.[5,6,7,8,17]

Key Finding 5: Multisystem End-Organ Effects of Chronic Hemodynamic Compromise

Hepatomegaly (5 cm below costal margin) reflected passive congestion from right heart failure with elevated central venous pressure.[9,14] Hepatic transaminase elevation (ALT 68, AST 82) indicated ischemia from reduced perfusion pressure.[9,14] Lethargy with altered sensorium resulted from combined hypoxemia (SpO2 88%), hypercapnia (pCO2 48), reduced cerebral perfusion from SVT (212 bpm), and metabolic acidosis (pH 7.28).[9,14] Rapid reversal of these findings with appropriate acute management (oxygen, adenosine, diuretics, antibiotics) demonstrated reversibility of end-organ effects.[9,14] This case illustrates how unoperated complex CHD produces multisystem involvement extending far beyond cardiac system, emphasizing urgency of definitive intervention.[9,11,15,18]

Conclusion

Unoperated complex congenital heart disease can achieve unexpected hemodynamic compensation into early adolescence, yet this compensation remains invariably precarious and subject to rapid, life-threatening decompensation when confronted with acute stressors such as respiratory infection and cardiac arrhythmia. This 12-year-old with an exceptionally rare triad (AVSD + dextrocardia + single atrium) survived to adolescence despite profound cardiac complexity—an unusual trajectory warranting investigation. Her acute presentation demonstrates how respiratory infection and recurrent SVT can rapidly unmask limited cardiac reserve in compensated complex disease. Standard acute management (adenosine for SVT termination, diuretics for CHF, oxygen, antibiotics) successfully reversed acute hemodynamic crisis, but such interventions cannot substitute for definitive repair. At age 12 with already-established moderate pulmonary hypertension (PA pressure 58 mmHg) and chronic cyanosis (baseline SpO2 85-88%), this patient is at critical temporal window before progression to irreversible pulmonary vascular obstructive disease. Urgent cardiac catheterization, electrophysiologic evaluation, and surgical consultation are essential. For clinicians managing unoperated complex CHD, this case underscores that: (1) acute respiratory infections and arrhythmias may precipitate life-threatening crises; (2) standard acute management remains effective even in unusual anatomy; and (3) definitive cardiac intervention must not be indefinitely deferred. Natural history of unoperated complex CHD typically progresses toward PVOD and reduced survival; timely definitive intervention offers best opportunity for improved survival and quality of life.

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