nificance of the clinical criteria of KD especially when “sup-
ported” by the finding of a dilated CA.

039

CONGENITAL HEART DISEASE CONFOUNDING THE
DIAGNOSIS OF ARRHYTHMOGENIC RIGHT VENTRICULAR
CARDIOMYOPATHY

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BACKGROUND: Early intervention in the diagnosis of arrhyth-
mogenic right ventricular cardiomyopathy (ARVC) can pre-
vent irreversible changes that may result in ventricular dys-
function and lead to sudden cardiac death. The 2010 revised
ARVC/D diagnostic Task Force Criteria (TFC) has in-
creased diagnostic sensitivity for ARVC while remaining
highly specific. However, certain conditions continue to
mimic ARVC and therefore warrant careful attention, espe-
cially when dealing with pediatric populations. We present
six cases in which clinical presentation has confounded the
diagnosis of congenital heart disease (CHD) versus ARVC.

METHODS: We reviewed medical charts for clinical, electrocardio-
graphic and imaging data. The results were anonymized, tabulated
and evaluated using the revised diagnostic criteria for ARVC.

RESULTS: We identified five patients in whom the presentation
of undiagnosed or seemingly unimportant CHD was initially
attributed to ARVC. RV or RVOT dilation was present in all,
with or without wall motion abnormalities. Filtered QRS was
prolonged in four cases. Significant ventricular extrasystole was
noted in two cases. Three of our cases demonstrated increased
Qp:Qs. The final diagnosis was most often partial anomalous
pulmonary venous connection (PAPVC). Despite an initial
high index of suspicion for ARVC, in all cases we eventually
attributed RV structural and functional changes, depolariza-
tion/conduction abnormalities and arrhythmias to left-to-right
shunts secondary to CHD. In contrast, we identified one pa-
tient in whom mild CHD masked the likely diagnosis of
ARVC. This patient presented with similar findings, includ-
ing: RV dilation, prolonged filtered QRS, significant ven-
tricular extrasystole and increased Qp:Qs. Despite closure
of the ASD, RV dilation persisted and she developed epi-
sodes of loss of consciousness, increasing suspicion of un-
derlying ARVC.

Table 1. Presentation and Diagnoses of Confounding Cases
* according to revised TFC

<table>
<thead>
<tr>
<th>Case</th>
<th>Age of presentation</th>
<th>Gender</th>
<th>Presenting Complaint</th>
<th>Initial Diagnosis</th>
<th>Major Criteria</th>
<th>Minor Criteria</th>
<th>ARVC Diagnosis by Revised Criteria</th>
<th>Final Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>11 years</td>
<td>male</td>
<td>palpitations</td>
<td>ARVC</td>
<td>I</td>
<td>IV, V</td>
<td>definite</td>
<td>PAPVC</td>
</tr>
<tr>
<td>2</td>
<td>12 years</td>
<td>male</td>
<td>syncope</td>
<td>ARVC</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>PAPVC</td>
</tr>
<tr>
<td>3</td>
<td>17 years</td>
<td>male</td>
<td>chest pain</td>
<td>ASD/ARVC</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>ASD</td>
</tr>
<tr>
<td>4</td>
<td>2 months</td>
<td>male</td>
<td>murmur</td>
<td>ARVC</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>PAPVC</td>
</tr>
<tr>
<td>5</td>
<td>6 years</td>
<td>female</td>
<td>palpitations</td>
<td>ASD</td>
<td>I</td>
<td>IV</td>
<td>borderline</td>
<td>ARVC</td>
</tr>
<tr>
<td>6</td>
<td>13 years</td>
<td>female</td>
<td>murmur, palpitations,</td>
<td>ARVC</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>PAPVC, ASD</td>
</tr>
</tbody>
</table>

CONCLUSION: ARVC poses a diagnostic challenge, especially
in pediatric populations where findings may overlap with
CHD. Cases of CHD with left-to-right shunting may lead
to structural and electrophysiologic findings consistent with
ARVC. In some cases, the findings are quite compelling and
may even meet the revised diagnostic criteria for ARVC.
On the other hand, underlying ARVC may co-exist with
and be masked by mild CHD. Despite a high index of sus-
picion for ARVC, careful echocardiography and MRI are
essential to detect covert CHD in the pediatric evaluation of
ARVC.

040

WITHDRAWN

041

NON-INVASIVE ASSESSMENT OF RIGHT HEART AND
PULMONARY VASCULAR COUPLING IN CHILDREN WITH
PULMONARY HYPERTENSIVE VASCULAR DISEASE: A
SIMULTANEOUS ECHOCARDIOGRAPHIC AND
CATHETERIZATION STUDY

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BACKGROUND: Cardiac catheterization is the gold standard
for assessment of hemodynamics in children with pulmo-
nary hypertensive vascular disease (PHVD). There is a need
for accurate, non-invasive correlates of these hemodynam-
ics. We aimed to identify correlations between echocardiographic
and catheter parameters in children undergoing card-
diac catheterization to investigate PHVD.

METHODS: Echocardiograms were performed on patients with
PHVD undergoing cardiac catheterization, after induction of
anesthesia. Echocardiographic parameters assessed included tr-
icuspid valve (TV) annular tissue Doppler velocities (TDD), TV
inflow Doppler, right atrial (RA) and right ventricular (RV)
dimensions and function. Cardiac catheterization data in-
cluded RA and RV pressures, pulmonary arterial pressure
(PAP), pulmonary blood flow, pulmonary vascular resistance
index (PVRI), pulmonary capacitance index (PCI) and cardiac
index (CI).

RESULTS: We studied 14 consecutive patients (8 male; median
age 6 years, range 1 - 15) with mean PAP 42 ± 22 mmHg and
PVRI 13 ± 6 WU/m². TV peak regurgitant velocity correlated
with systolic PAP (r=0.79, p<0.01) suggesting patients were
studied under the same hemodynamic conditions. RA mean
pressure correlated with TV E/e prime ratio (r=0.67, p=0.02).
There was no correlation between echocardiographic parameters
of RV function (TAPSE, MPI, TV S prime) and
catheter parameters. PVRI correlated with TV TDI a prime
(r=0.56, p=0.03). CI correlated with TV inflow E velocity