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## BACKGROUND

- Heart Valve Centers of Excellence are well-described in adults, however not in the pediatric population.
- Congenital and acquired valve diseases are some of the commonest and most severe forms of cardiac disease in pediatrics, and guidelines are lacking.
- Infants and children have valvular pathology that is distinct from that of adults, as well as surgical considerations that vary based on age, size, growth potential, and lifestyle.
- The Pediatric Heart Valve Center at the Children's Hospital of Philadelphia (CHOP) was established to fill the role of a center of excellence in pediatric valve disease.

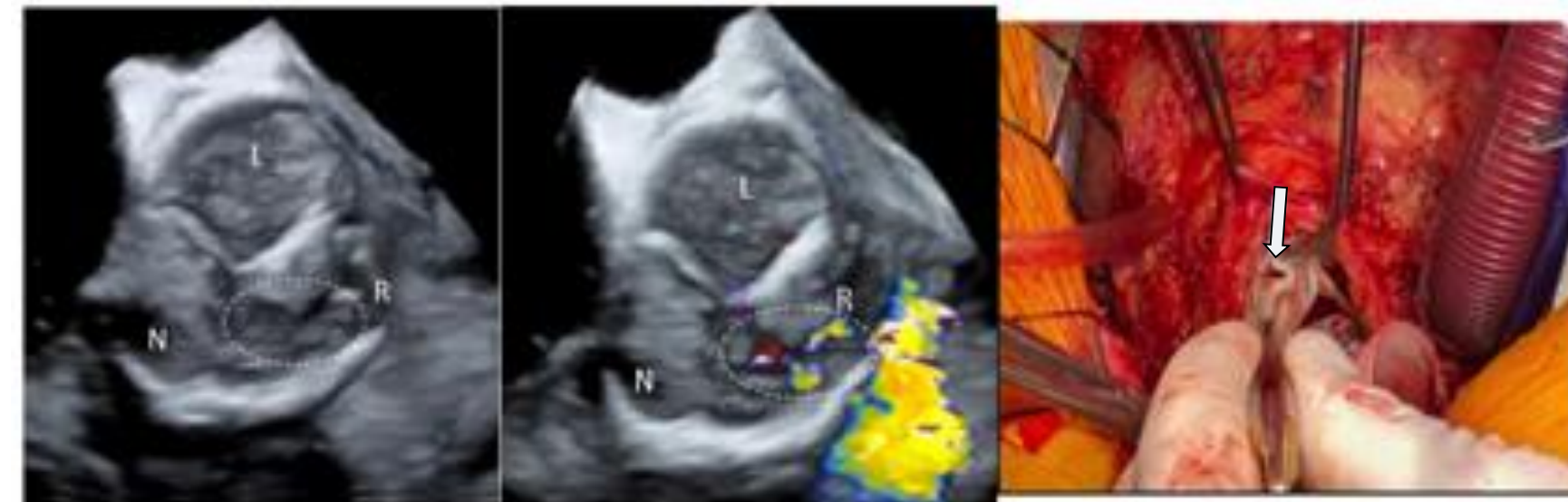
## PROGRAM STRUCTURE

- Small, multi-disciplinary group of echo-trained cardiologists, cardiothoracic surgeons, cardiac interventionalist, cardiac anesthesia, cardiac sonographers, and advanced cardiac imaging trainees.
- Grant funding received from CHOP Frontier program
- At inception, the team was kept small, ensuring highly engaged individuals who were allotted dedicated time to focus on valve center initiatives and skill development.
- Dedicated program coordinator and research nurse work to manage referral volume, coordinate clinical information/scheduling, facilitate meetings, and maintain record of clinical recommendations and outcomes.
- Biweekly patient review meetings allow discussion of referred cases, with recommendations for further imaging, need for intervention, and options for surgical intervention decided; monthly educational meetings allow team to re-review cases, discuss learning experiences, and advance knowledge of relevant topics.
- Ongoing collaboration with clinical research arm, including 3D modeling from both 3D echo and cross-sectional imaging for surgical planning.

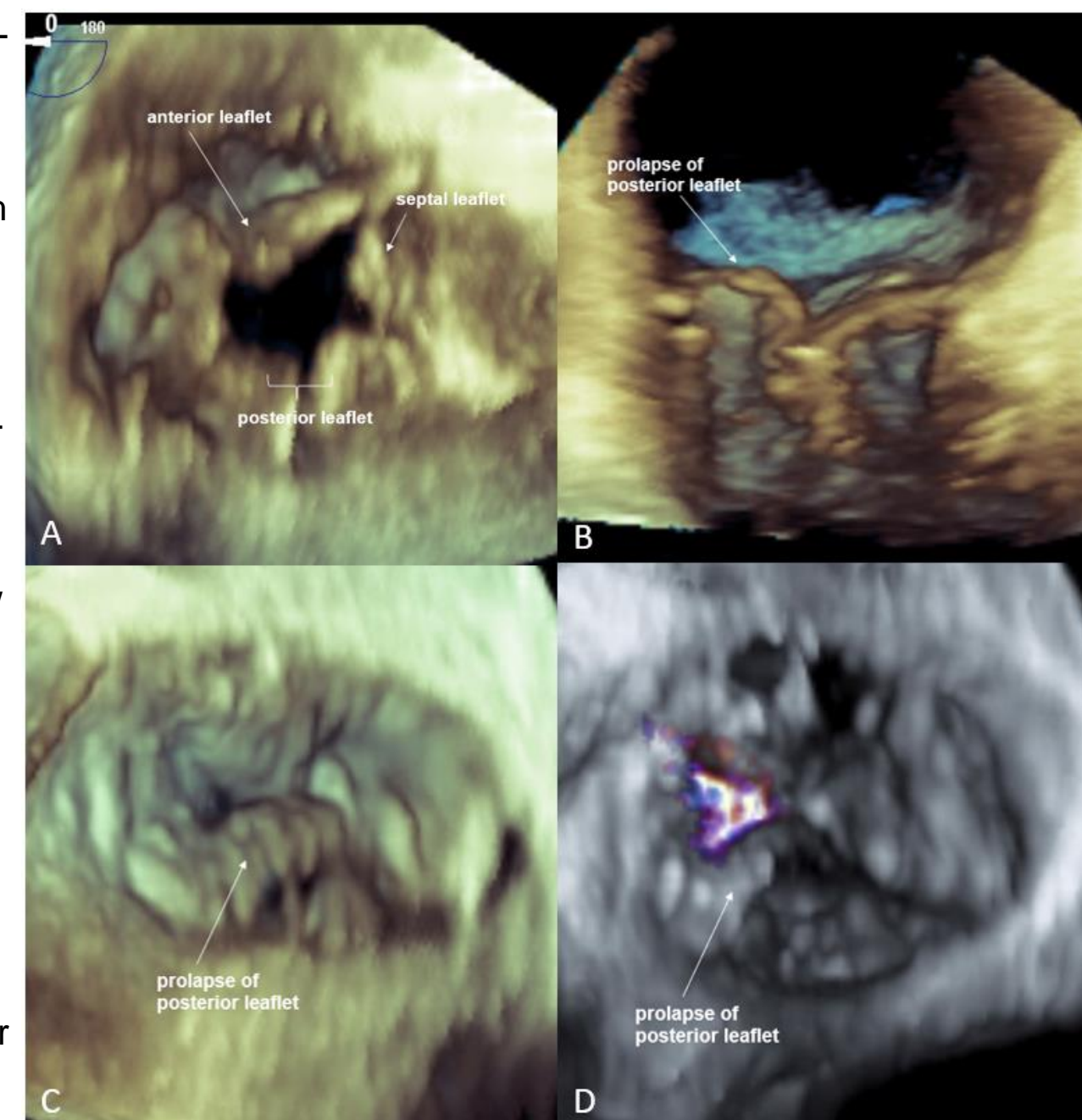
## EARLY CASES AND IMAGING

- 87 patients referred to the Pediatric Heart Valve Center since September 2020 were reviewed
- 24 had surgery following a valve center discussion
- Types of valve intervention included:
  - 8 aortic valve
  - 4 truncal valve
  - 4 mitral valve
  - 3 neo-aortic valve
  - 4 common AV valve
  - 1 tricuspid valve
- Valve center consult notes and preoperative echo reports were reviewed & compared with operative notes (Table 1)

**Figure 1.** Left: 3D TEE images demonstrating a perforated right coronary cusp (left, dashed circle) with insufficiency originating through the perforation (middle, dashed circle). Findings were confirmed intraoperatively (right, arrow).



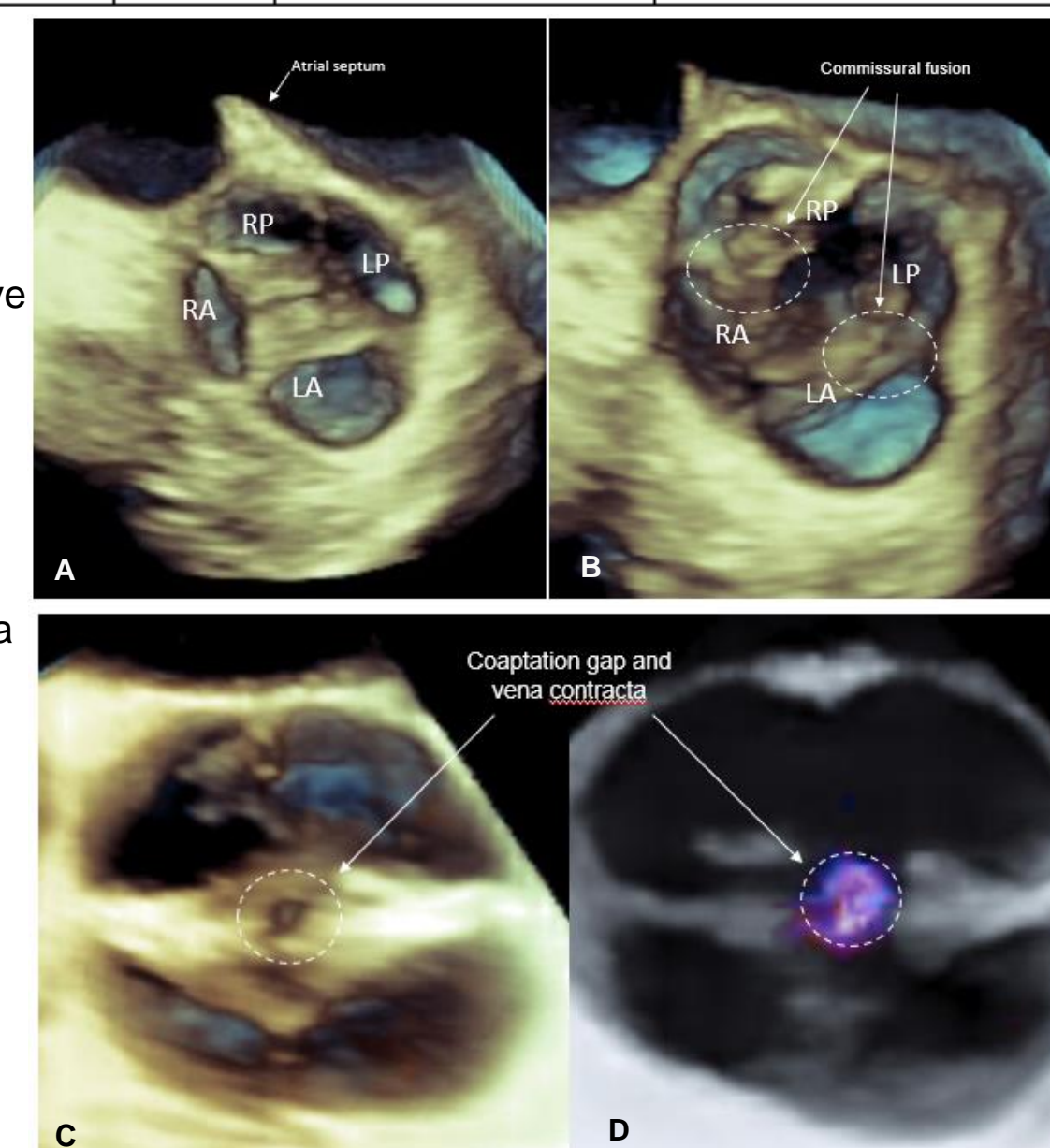
**Figure 2.** 3D TEE, mid-esophageal 0° view of tricuspid valve in HLHS; patient B. (A) open valve enface from ventricle looking up, with a notably deficient septal leaflet, (B) prolapse of the posterior leaflet from 4-chamber view, (C) enface view of the tricuspid valve in systole; surgeon's view from atrium. There is significant prolapse of the posterior valve leaflet, (D) same view with color, the jet of regurgitation originates from the anterior/posterior coaptation gap along the prolapsing posterior leaflet.



**Table 1.** Preoperative and intraoperative findings in a random, representative sample of valve center patients

	Medical History	Age, sex	Valve Center description based on preoperative echo findings	Intraoperative findings
A	Tetralogy of Fallot status post repair with progressive aortic insufficiency	12 yr, F	Trileaflet valve, R coronary cusp with perforation near annulus	Perforation at base of right coronary cusp (Figure 1)
B	Hypoplastic left heart syndrome palliated to lateral tunnel Fontan with moderate to severe tricuspid regurgitation	14 yr, M	Prolapse of anterior and posterior leaflets, shortened septal leaflet	Posterior leaflet redundant, dysplastic, and prolapsing; diminutive septal leaflet; anterior leaflet appeared largely normal (Figure 2)
C	Truncus arteriosus status post repair with truncal valve insufficiency	9 yr, F	Quadracuspid valve, fusion of right/left and between two "non" commissures, thick/rolled leaflets	Quadracuspid valve, fusion of right anterior/posterior and left anterior/posterior leaflets, thickened valve (Figure 3)
D	Trisomy 21, VSD, cleft mitral valve, pulmonary stenosis status post patch VSD closure, & RVOT augmentation with progressive mitral regurgitation	16 mo, F	Prolapse of anterior leaflet (A2, some A3), appearance of mitral valve cleft may be due to prolapse, not true cleft	Anterior leaflet redundant and prolapsed along A1/A2, posterior leaflet appeared usual, no mitral valve cleft
E	Progressive aortic insufficiency	14 yr, F	Trileaflet valve, noncoronary cusp deficient, abnormal motion, prolapse; right and left cusps normal	Perforation of noncoronary leaflet
F	Mitral valve prolapse with severe mitral regurgitation	4 yr, M	Prolapse of anterior leaflet, restrictive motion of posterior leaflet	Dysplastic valve, severe prolapse of A2, chordal support to A1 fibrotic

**Figure 3.** 3D TEE images demonstrating an enface view of the truncal valve in patient A. The valve is quadracuspid. With valve opening, fusion of the right anterior/posterior and left anterior/posterior commissures is observed (A, B). With valve closure, there is a central coaptation defect through which the jet of insufficiency originates (C, D). RA=right anterior, RP=right posterior, LA=left anterior, LP=left posterior



## FINDINGS

- A dedicated, multidisciplinary team has allowed our group to build skills and experience in evaluation and treatment of pediatric heart valve disease. Lessons learned include:
- Sedation performed by an anesthesiologist well-versed in 3D acquisition breath holds and positioning is important for quality of image acquisition
- 3D imaging, including TEE, in advance of OR allows more time for cropping, interpretation, and multidisciplinary discussion
- Educational conferences allow for review of current practices and team-initiated improvement
- Partnering with clinical research colleagues has been mutually beneficial and has advanced clinical applications
- Collaboration with invested surgeons is crucial to refining imaging techniques & expanding surgical options for patients

## CONCLUSIONS & FUTURE DIRECTIONS

- Creation of a Pediatric Heart Valve Center has allowed for improved collaboration and surgical planning in pediatric valve disease.
- Continued efforts will refine valve imaging protocols and enable streamlined discussions regarding valve intervention, with the goal of improving outcomes.
- Increased collaboration with cross-sectional imaging colleagues will allow for creation of CT/MRI protocols for valve imaging.
- Research efforts in interventional techniques, surgical approaches, and advanced imaging including 4D will continue to improve our management of pediatric valve disease.

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