DOI: 10.7860/JCDR/2026/87784.24521

Review Article

Surgery Section

Pentalogy of Cantrell: A Narrative Review of Aetiopathogenesis, Diagnostic Advances and Contemporary Management Strategies

SHAILABH[1], IMRAN ALI KHAN[2], ANUP ZADE[3], YASHASVI TRIVEDI[4], BHAGYESH SAPKALE[5]

ABSTRACT

Pentalogy of Cantrell (POC) is known as an extremely rare congenital disorder which is characterised by a distinctive constellation of five midline defects involving anterior abdominal wall, lower sternum, anterior diaphragm, diaphragmatic pericardium and heart, often accompanied by complex intracardiac anomalies. The clinical spectrum of POC usually ranges from complete forms exhibiting all five defects to incomplete type having partial expression thus contributing towards significant heterogeneity in presentation and outcomes. The present narrative review article summarises current understanding of embryologic and molecular basis of POC thereby highlighting the role of early mesodermal developmental failure during the first weeks of gestation. Advances in prenatal diagnosis using ultrasonography, foetal echocardiography, Three-Dimensional (3D) imaging as well as foetal magnetic resonance imaging have improved early detection, anatomical delineation along with prognostic assessment. Postnatal imaging supports into comprehensive evaluation and surgical planning of POC cases. Management of POC thus remains primarily surgical while it also needs a coordinated multidisciplinary approach inclusive of prenatal counselling, neonatal intensive care, cardiology and paediatric surgery. Both single-stage as well as staged surgical strategies are used based upon disease severity, patient stability along with emerging perinatal and foetal interventions offering potential benefits in selected cases. Despite improvements in diagnostic as well as therapeutic modalities, prognosis remains variable which is closely linked to the severity of intracardiac and thoraco-abdominal defects. Continued research focusing on genetic mechanisms, long-term outcomes, standardised management protocols is said to be an essential aspect to optimise care and improve survival, quality of life in patients having POC.

Keywords: Anterior abdominal wall defect, Cardiac malformations, Mesodermal development, Prenatal imaging, Thoraco-abdominal anomalies

INTRODUCTION

The POC is known to be an extremely rare congenital disorder which is characterised by a distinctive combination of five midline defects affecting the lower sternum, anterior diaphragm, diaphragmatic pericardium, supra-umbilical abdominal wall and the heart with associated intracardiac anomalies [1]. The clinical expression of POC exists along a wide spectrum that ranges from complete forms having all five defects to partial variants which show only few components [2]. These malformations arise due to a disruption in early embryonic development usually resulting from defective formation, differentiation and migration of mesodermal structures during approximately 14-18 days of embryonic life [1,3]. This early mesodermal disruption further impairs development of transverse septum of the diaphragm, ventral body wall resulting into the characteristic thoraco-abdominal anomalies [3]. POC is usually associated with ectopia cordis as well as supra-umbilical omphalocele which contribute to clinical severity and prognosis [4,5]. The spectrum of associated intracardiac anomalies is broad inclusive of ventricular septal defect, atrial septal defect, tetralogy of Fallot, other complex congenital heart diseases, which are key determinants of survival of patients having POC [4,6].

The POC was properly described by Cantrell, Haller, Ravitch in 1958, who reported a series of patients showing this unique constellation of defects thereby it helped into establishing it as a recognisable clinical syndrome [7]. Population-based estimates suggest an incidence which ranges from approximately 5.5-7.9 per million live births while prevalence reported is 1 in 65,000 to 200,000 live births [1,8]. The POC appears to show a slight male predominance having reported male-to-female ratios of about 1.3-1.4:1 [8,9]. Geographic distribution does not show a clear region-wise clustering although a larger proportion of reported cases originate from Europe, North America usually reflecting differences in diagnostic access as well as reporting practices [9]. As many of the severe cases are detected prenatally it can result in foetal demise, termination of pregnancy thereby true incidence can be underestimated in live-birth statistics [9].

Toyama later proposed a classification system in year 1972 for adequately stratifying the syndrome based on the presence, completeness of the associated defects thus aiding in diagnosis and management planning [10]. The present narrative review article further aimed to highlight about aetiopathogenesis, diagnosis, management along with outcomes of POC to support informed multidisciplinary clinical care.

Classification of Pentalogy of Cantrell (POC)

The POC is usually characterised by a specific constellation of midline defects inclusive of anterior abdominal wall, lower sternum, anterior diaphragm, diaphragmatic pericardium and intracardiac structures [1,11]. Although classic description includes all five anomalies, clinical presentations can further significantly vary in number, combination of defects observed. Toyama's classification widely helps clinicians to categorise degree of expression of syndrome [10]. Class I (definite or complete) are cases in which all five characteristic defects are present [11]. Class II (probable) which refers to presentations with four of five defects, usually including both ventral wall and intracardiac anomalies [10,12]. Class III (incomplete) is of cases having various combinations of anomalies, usually with a sternal defect but not fulfilling the criteria for a probable or complete diagnosis [10,11].

The clinical usage of such a classification lies in its ability for guiding prognosis and management, as the presence or absence of specific defects especially major intracardiac lesions mostly

influences surgical planning, outcomes [11]. Complete forms (Class I) is inclusive of a higher risk, complex need of surgical intervention. Probable forms (Class II) require comprehensive evaluation as well as staged surgical intervention [11,12]. The incomplete forms (Class III) can usually present having less severe combined anomalies but it requires individualised care [11]. By properly classifying patients according to number, severity of anomalies present, Toyama's classification thereby provides a practical framework for clinicians confronting POC [11]. Toyama's Classification of POC is described in [Table/Fig-1] [10-12].

Class	Terminology	Defining features	Clinical implications	Reference(s)
Class I	Definite / complete	All five defects: anterior abdominal wall, lower sternum, anterior diaphragm, diaphragmatic pericardium, intracardiac anomalies	Highest severity and surgical complexity; often requires early, complex intervention	[10,11]
Class II	Probable	Four of the five defects, typically including ventral abdominal wall and intracardiac anomalies	Requires comprehensive evaluation and often staged surgical management	[10,12]
Class III	Incomplete	Variable combination of defects, usually including a sternal defect, but not meeting criteria for complete or probable forms	Generally less severe; individualised management based on specific anomalies	[10,11]

[Table/Fig-1]: Toyama's Classification of Pentalogy of Cantrell (POC) [10-12]. Made by Authors

Clinical Features of Pentalogy of Cantrell (POC)

The POC manifests as a distinctive constellation of congenital anomalies which all together define its clinical phenotype [13]. POC is inclusive of midline thoraco-abdominal defects such as a supra-umbilical anterior abdominal wall defect such as omphalocele, which often allows protrusion of abdominal viscera covered by a membrane [13]. Associated with this is a defect in the lower sternum which ranges from a sternal cleft to complete absence thereby predisposing to ectopia cordis, where heart is partially or entirely located outside thoracic cavity [14]. Additional structural deficits include deficiency of anterior diaphragm as well as diaphragmatic pericardium resulting into abnormal communication between body cavities and herniation of organs into the thoracic space [15,16].

Central to clinical presentation of POC are intracardiac anomalies, which are highly variable but mostly documented in all the affected cases [15]. The most frequent cardiac lesions are inclusive of ventricular septal defects, atrial septal defects having more complex malformations such as tetralogy of Fallot, double-outlet right ventricle and ventricular diverticulum [15]. These cardiac anomalies usually produce cardiac murmurs, cyanosis, haemodynamic instability shortly after birth as well as significantly influence prognosis, management strategies [14,15]. POC can be associated further having a range of extracardiac anomalies which are craniofacial malformations, limb abnormalities and other visceral defects, contributing to its broad phenotypic spectrum also the clinical challenges in diagnosis, care [13,17].

Rare and Atypical Clinical Manifestations of Pentalogy of Cantrell (POC)

The POC can also present showing rare, unusual clinical features which extend beyond its classic pentad of midline defects [18]. In some of the reported cases, POC has been associated having additional structural anomalies not typically ascribed to syndrome, thereby reflecting its broad phenotypic variability [18]. For example, there are few documented instances of POC occurring along with craniosynostosis, hypogenesis of the corpus callosum as well as tonsillar cerebellar herniation thus indicating involvement of central nervous system in addition to thoraco-abdominal malformations [18]. Such features highlight potential for inclusiveness of complex multisystem in affected individuals which underscores about the importance of adequate imaging as well as multidisciplinary assessment [18].

Beyond structural anomalies of brain-skull, rare associations such as single umbilical artery, other vascular anomalies which can be usually identified on detailed foetal scans, postnatal evaluation thereby further suggesting deviations in vascular development alongside the primary defects [19]. Additionally, unusual combinations inclusive of gastroschisis with cystic hygroma have been reported in early gestation thus illustrating that POC can present with anomalies of the lymphatic, abdominal wall systems which is not known to the part of canonical pentad [20]. All of these findings while infrequent, reflect embryologic complexity as well as variability of POC and support the need for comprehensive prenatal imaging, genetic evaluation [19,20].

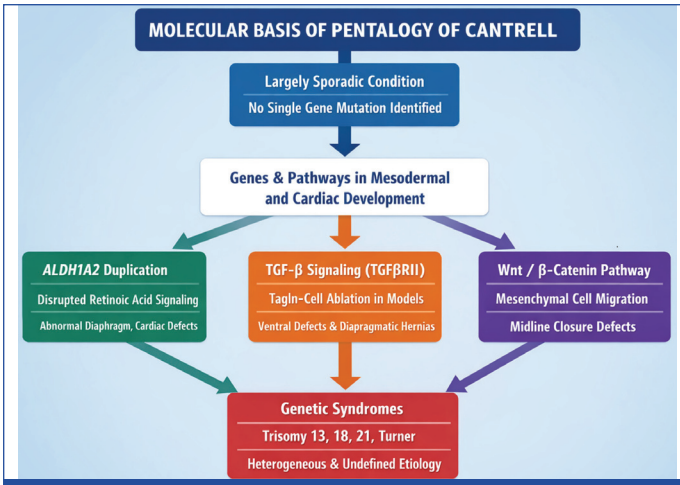
Pathogenesis and Molecular Basis of Pentalogy of Cantrell (POC)

The POC is believed to arise from an early embryologic failure in ventral midline development usually involving defective differentiation, proliferation as well as migration of lateral mesodermal and ventral body wall structures during the first 14-18 days of embryogenesis [21]. This disruption further results into a spectrum of midline closure defects which affect abdominal wall, lower sternum, anterior diaphragm, diaphragmatic pericardium and heart [21]. The concept of a ventral midline "developmental field" further reflects that these anomalies cluster due to perturbation of a common embryonic region where adjacent mesodermal elements normally fuse and differentiate thereby explaining about frequent associations with other midline anomalies (e.g., cleft lip/palate) seen in cases of POC [21].

Considering the molecular level, no single gene mutation has been identified definitively as universal cause of POC also the condition is largely sporadic in nature [3]. However, case reports as well as experimental models suggest roles for genes and pathways important in mesodermal, cardiac development [3,22]. For instance, duplication of ALDH1A2 which is a gene encoding enzyme that produces retinoic acid (a critical morphogen in early embryogenesis), has been reported in association with syndrome, implying disrupted retinoic acid signalling can contribute to abnormal diaphragm, cardiac morphogenesis [3,23]. Experimental ablation of Transforming Growth Factor-beta (TGFβ) signalling (TGFβRII) in Tagln-expressing cells in model organisms produces ventral midline closure defects along with diaphragmatic hernias which further resembles features of POC thus highlighting the importance of mesenchymal signalling pathways in ventral body wall formation [22]. Other developmental pathways inclusive of Wnt/β-catenin which is involved in mesenchymal cell migration have also been implicated in midline defects relevant to POC [1,13]. Although the genetic syndromes (such as trisomies 13, 18, 21, Turner) show occasional association, precise molecular aetiology remains heterogeneous and incompletely defined thus highlighting further about an unmet need for further genetic and developmental studies [4,24]. Molecular basis of POC is depicted through [Table/Fig-2].

Genetic Aspects of Pentalogy of Cantrell (POC)

Although POC is usually considered a sporadic condition, several observations also suggest a potential genetic contribution into its development [25]. Familial occurrences have been reported in rare cases thereby raising possibility of inherited susceptibility [25]. Some investigators have proposed an X-linked pattern of inheritance usually in male-predominant cases having thoraco-



[Table/Fig-2]: Molecular basis of Pentalogy of Cantrell (POC).
Made by Authors

abdominal midline defects and ectopia cordis [25]. Additionally, associations with chromosomal abnormalities have been described inclusive of trisomy 13, trisomy 18 and Turner syndrome thus supporting hypothesis that chromosomal imbalance during early embryogenesis can predispose to POC [26]. Advances in prenatal genetic testing including karyotyping, chromosomal microarray analysis have therefore been recommended when POC is suspected on foetal imaging for detection of underlying genetic abnormalities to guide parental counselling [23]. However, due to rarity of the condition as well as limited large-scale genetic studies, exact inheritance pattern, genetic determinants of POC remain incompletely understood thus warranting further investigation using modern genomic approaches.

Diagnostic Approach and Imaging Modalities in Pentalogy of Cantrell (POC)

The POC is a rare congenital syndrome which is defined by a characteristic combination of five midline anomalies inclusive of a supra-umbilical abdominal wall defect (often omphalocele), lower sternal defect, anterior diaphragmatic defect, deficiency of the diaphragmatic pericardium and congenital intracardiac malformations such as ventricular septal defect or tetralogy of Fallot [27,28]. These features result from aberrant embryological mesodermal development which can be present in complete or partial forms along with complete presentations including all five defects [27]. Early and accurate diagnosis is very essential because of the severity of the anomalies usually the intracardiac and thoraco-abdominal defects which further heavily influences prognosis and management planning of POC cases [29].

Diagnosis of POC starts prenatally with the use of routine obstetric imaging [30]. Two-Dimensional (2D) ultrasound is main aspect of antenatal detection, which helps to identify hallmark anterior wall as well as thoracic abnormalities including herniation of abdominal contents, ectopia cordis (heart located outside the thoracic cavity) [30,31]. Foetal echocardiography complements this through delineating associated cardiac defects which are important for counselling, surgical planning of the POC patients [26]. When additional details are required, advanced imaging like foetal MRI and postnatal Computed Tomography (CT) can further help for clarifying extent of thoracic, diaphragmatic as well as pericardial defects [32,33]. After birth, echocardiography, CT and MRI are mostly used in combination for confirmation of the full spectrum of anomalies while also it helps in multidisciplinary surgical strategy [32,34]. Diagnostic approach in POC is described in [Table/Fig-3] [26,31-34].

Emerging Diagnostic Modalities in Pentalogy of Cantrell (POC)

Emerging approaches into diagnosis of POC have focused on enhancing early detection accuracy as well as comprehensive anatomical delineation beyond conventional 2D imaging [35]. Although case reports remain predominant source of evidence, 3D ultrasound along with advanced foetal imaging techniques have been reported for improvement of structural characterisation of complex ventral-thoracic anomalies that are associated with POC [35]. A 3D sonographic assessment in prenatal settings further allows volumetric visualisation of anterior thoraco-abdominal wall as well as associated malformations potentially facilitating clearer differentiation of ectopia cordis, midline defects when compared to traditional 2D scans alone [36]. There are documented cases in which emphasise about 3D ultrasound utility in both third-trimester, early pregnancy assessments of POC thereby highlighting its value in revealing spatial relationships of anatomical abnormalities [36,37].

Foetal MRI has been increasingly used as a complementary modality which enhances diagnostic precision for POC [38]. MRI provides superior soft-tissue contrast as well as multiplanar capabilities which can better define extent of diaphragmatic and thoraco-abdominal defects [38]. MRI further provides appropriate information about the relation of heart to surrounding structures usually in cases of ectopia cordis or complex abdominal wall involvement that may be challenging on ultrasound alone [38,39]. The combined usage of foetal echocardiography along with MRI can be helpful for optimisation of prenatal evaluation, offering improved delineation of both cardiac and non cardiac components of syndrome as well as valuable prognostic information for multidisciplinary planning in POC cases [1,38].

Treatment and Management of Pentalogy of Cantrell (POC)

Initial Management

The management of POC is usually surgical and multidisciplinary which is focused on correcting complex combination of anomalies

Stage of diagnosis	Imaging modality	Key findings identified	Clinical utility	Reference(s)
Prenatal	Two-Dimensional (2D) ultrasonography	Omphalocele, ectopia cordis, anterior abdominal wall and sternal defects	Primary screening tool for early detection and suspicion of POC	[30,31]
Prenatal	Foetal echocardiography	Ventricular septal defect, atrial septal defect, tetralogy of Fallot, other intracardiac anomalies	Defines cardiac anatomy, aids prognostication and parental counselling	[26]
Prenatal (adjunct)	Foetal Magnetic Resonance Imaging (MRI)	Detailed visualisation of thoraco-abdominal structures, diaphragm, pericardium	Improves anatomical assessment when ultrasound findings are limited	[32,33]
Postnatal	Transthoracic echocardiography	Confirmation of intracardiac defects, evaluation of cardiac function	Guides surgical planning and perioperative management	[32,34]
Postnatal	Computed Tomography (CT)	Sternal defects, thoracic cage abnormalities, diaphragmatic discontinuity	Provides precise anatomical detail for reconstructive surgery	[32,34]
Postnatal (adjunct)	Magnetic Resonance Imaging (MRI)	Soft-tissue relationships, mediastinal anatomy, pericardial defects	Complements CT while avoiding ionising radiation	[32-34]

[Table/Fig-3]: Diagnostic approach in Pentalogy of Cantrell (POC) [26,31-34].

inclusive of the heart, diaphragm, sternum, pericardium, anterior abdominal wall [40]. The definitive treatment usually involves careful planning with the help of a multidisciplinary team which spans around prenatal counselling, neonatal intensive care, cardiology as well as paediatric surgical specialties [40]. The overarching goals of surgical intervention are firstly inclusive of correcting or palliating intracardiac malformations, secondly to restore heart to its anatomical position within mediastinum and thirdly to repair thoraco-abdominal as well as diaphragmatic defects for protection of heart and other internal organs [41,42]. Early surgical repair can be technically demanding which also carries significant risk; neonatal procedures are associated to higher mortality when compared with delayed approaches, as critical structures are small and thoracic cavity is also limited in space [43]. Therefore, stable infants can benefit from initial conservative management which can further allow for some physiologic maturation before proceeding forward for a definitive reconstruction, whereas critically-ill neonates require timely surgical intervention for preventing life-threatening complications associated with POC [43].

Surgical Repair

In a study of eight patients undergoing radical one-stage repair, multidisciplinary reconstruction of cardiac lesions as well as thoraco-abdominal defects achieved satisfactory mid-term outcomes without having any perioperative mortality thereby emphasising about importance of achieving adequate mediastinal space as well as securing the chest wall protection [44]. Staged management inclusive of initial repair of omphalocele and coverage of heart in the neonatal period along with delayed complete intracardiac corrective surgery later in performed in the childhood, also highlights that timing and sequencing of interventions are dictated by clinical status and complexity of anomalies [45]. Adjunctive perioperative considerations such as mesh or Gore-Tex reconstruction for abdominal wall defects, intra-abdominal pressure monitoring along with vigilant postoperative care owing to risks of respiratory insufficiency or cardiac dysfunction are useful in POC [46]. Overall, surgical repair remains mainstay of treatment, individualised, multidisciplinary planning which is based upon defect severity as well as stability of patient is an essential aspect to optimise outcomes in this rare and challenging condition [45,46].

Current Treatment Modalities

Current treatment modalities for POC emphasise early prenatal diagnosis, risk stratification as well as coordinated multidisciplinary management in tertiary care centres [47,48]. Advances in foetal imaging usually detailed foetal echocardiography, high-resolution ultrasonography allow proper characterisation of intracardiac defects and associated thoraco-abdominal anomalies thereby facilitating individualised perinatal planning [35,48]. Delivery is usually recommended in specialised centres equipped with neonatal intensive care, paediatric cardiac surgery along with advanced cardiopulmonary support [9,21]. Postnatal management usually integrates staged surgical reconstruction along with optimised perioperative critical care which is inclusive of ventilatory support, haemodynamic stabilisation and careful monitoring of cardiopulmonary function [16,43]. The individualised treatment algorithms based on the severity of cardiac anomalies, presence of ectopia cordis and size of abdominal wall defects are important aspects as these factors significantly influence surgical feasibility as well as overall prognosis of POC cases [30,43]. Such integrated treatment strategies have contributed into gradual improvement in survival, functional outcomes in selected patients having this rare congenital syndrome [43].

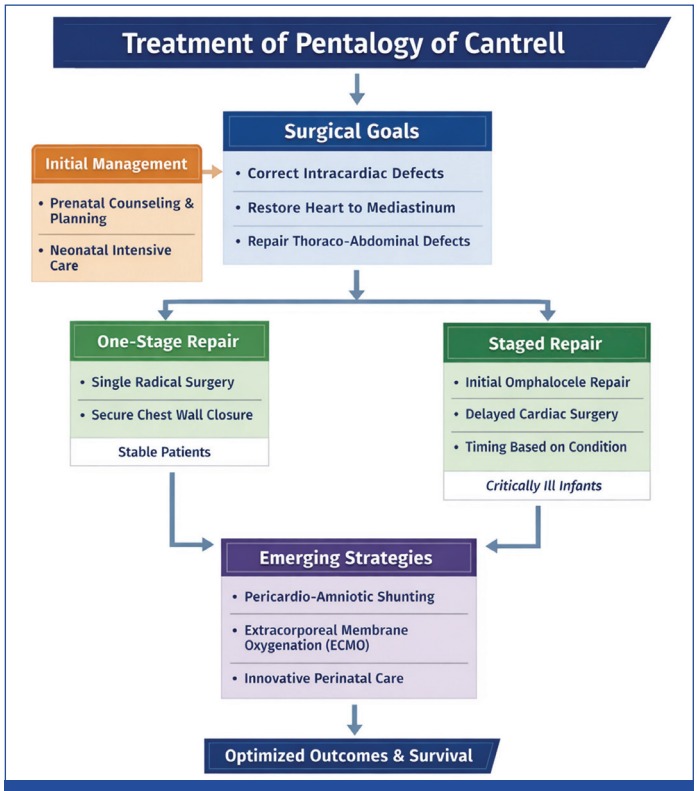
Emerging Management for POC

Emerging approaches into management of POC are gradually moving beyond traditional postnatal surgical repair toward innovative perinatal as well as multidisciplinary strategies which further aim to improve perinatal outcomes usually in severe or incomplete forms of the condition [49]. An emerging approach is pericardio-amniotic shunting which is performed prenatally in foetuses having large pericardial effusions associated with POC [49]. In a case reported, shunting of the pericardial fluid into amniotic cavity at around 33 weeks' gestation relieved lung compression, improved lung development also contributed to a more favourable neonatal course thereby allowing successful postnatal corrective surgery, eventual discharge home in good condition [49]. This foetal intervention strategy represents a novel way for addressing secondary physiological complications in utero which could otherwise compromise survival while also exacerbate pulmonary hypoplasia after birth [49].

Multidisciplinary perinatal care highlights about coordinated management which spans from advanced prenatal imaging, counselling to innovative postnatal support modalities, such as usage of Extracorporeal Membrane Oxygenation (ECMO), targeted ventilation strategies (e.g., inhaled nitric oxide) as well as individualised staged surgical sequencing which is guided by comprehensive risk assessment [16,50]. These approaches further seek into optimisation of physiological stability before, during and after definitive repair thus potentially reducing morbidity while also improving survival in patients [50]. The most emerging strategies remain grounded in case-based evidence which further requires systematic research, early foetal intervention as well as tailored, multidisciplinary perinatal protocols reflect a shift toward more proactive, physiology-based management in selected cases of high-risk foetuses and newborns having POC [50,51]. Treatment of POC is depicted in [Table/Fig-4].

Short and Long-Term Outcomes in POC

The prognosis of POC usually depends upon severity of associated cardiac anomalies, presence of ectopia cordis and the completeness of syndrome [1]. Historically, the survival rates in patients were reported to be approximately 30-40% where many affected neonates die within early postnatal period because of severe cardiac malformations, respiratory insufficiency, haemodynamic instability [1,4]. Outcomes are poor in the complete form of syndrome and in cases which are associated having significant ectopia cordis



[Table/Fig-4]: Treatment of Pentalogy of Cantrell (POC).
Made by Authors.

or complex intracardiac defects [52]. Nevertheless, advances in prenatal diagnosis, neonatal intensive care and staged surgical reconstruction have led to gradual improvement into survival in selected POC patients [21,43]. Among survivors, postoperative recovery can be prolonged which is frequently complicated by the need for extended ventilatory support as well as management of cardiopulmonary complications [26]. Long-term survivors of POC cases can achieve acceptable growth as well as functional outcomes after adequate surgical correction in patients [26,53]. However, it usually requires cardiology follow-up along with multidisciplinary care due to residual cardiac defects, complications related to thoraco-abdominal reconstruction [3,16].

Future Directions and Research Priorities in Pentalogy of Cantrell (POC)

Future research on POC must to be focused on better understanding underlying genetic, developmental causes of such a rare midline defect, as its exact aetiology remains unclear [1]. The genetic contributions and embryologic mechanisms inclusive of associations with chromosomal abnormalities as well as potential gene mutations could help clarify why POC occurs and how different phenotypes arise which can further ultimately inform targeted prenatal screening and counselling strategies [47]. Future investigations are likely to explore refinements in prenatal diagnostics, long-term outcomes as well as standardised management protocols with usage of larger multicenter cohorts and registries [47]. As POC exhibits wide variation in severity and outcomes, comprehensive data collection could thereby support evidence-based guidelines for risk stratification, timing along with sequencing of interventions, assessment of quality-of-life measures in survivors of POC [52]. This research direction aims for moving beyond isolated case reports toward systematic studies which can improve prognostication, therapeutic decision-making also family counselling [47,52].

CONCLUSION(S)

The POC is known as a rare, complex congenital syndrome which is characterised by a wide spectrum of midline defects along with significant variability in presentation, management and outcomes. Early, accurate prenatal diagnosis that is aided by advanced imaging modalities is very essential for optimal counselling as well as treatment planning. Management of POC further needs individualised, multidisciplinary approach which integrates surgical expertise along with an intensive perioperative care. Emerging prenatal, perinatal interventions show promise in improving outcomes while future research focusing on genetic mechanisms, standardised care protocols is very crucial for enhancing prognostication and long-term quality of life in cases of patients having POC.

REFERENCES

[1] Williams AP, Marayati R, Beierle EA. Pentalogy of Cantrell. Semin Pediatr Surg. 2019;28(2):106–10. Doi: 10.1053/j.sempedsurg.2019.04.006 PubMed PMID: 31072457; PubMed Central PMCID: PMC6559797.

[2] Kylat RI. Complete and incomplete pentalogy of Cantrell. Child (Basel). 2019;6(10):109. Doi: 10.3390/children6100109 PubMed PMID: 31590448; PubMed Central PMCID: PMC6826365.

[3] Jnah AJ, Newberry DM, England A. Pentalogy of Cantrell: Case report with review of the literature. Adv Neonatal Care. 2015;15(4):261-68. Doi: 10.1097/ANC.0000000000000209 PubMed PMID: 26225594.

[4] Sana MK, Rentea RM. Pentalogy of Cantrell. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2025 [cited 2026 Jan 14]. Available from: http://www.ncbi.nlm.nih.gov/books/NBK558948/ PubMed PMID: 32644374.

[5] Fazea M, Alhameli M, Ahmed F, Askarpour MR, Murshed W, Jarwsh A, et al. Pentalogy of Cantrell associated with ectopia cordis: A case report. Pediatr Health Med Ther. 2022;13:283-87. Doi: 10.2147/PHMT.S374289 PubMed PMID: 35996555; PubMed Central PMCID: PMC9391988.

[6] Hill MA, Kwon JH, Rajab TK, Kavarana MN. Pentalogy of Cantrell with a left ventricular diverticulum and muscular VSD. Ann Thorac Surg. 2022;113(6):e461-e463. Doi: 10.1016/j.athoracsur.2021.07.093 PubMed PMID: 34481798.

[7] Cantrell JR, Haller JA, Ravitch MM. A syndrome of congenital defects involving the abdominal wall, sternum, diaphragm, pericardium, and heart. Surg Gynecol Obstet. 1958;107(5):602-14. PubMed PMID: 13592660.

[8] Zvizdic Z, Sefic-Pasic I, Mesic A, Terzic S, Vranic S. The complete spectrum of pentalogy of Cantrell in one of a set of dizygotic twins: A case report of a rare congenital anomaly. Medicine (Baltimore). 2021;100(14):e25470. Doi: 10.1097/MD.0000000000025470 PubMed PMID: 33832161; PubMed Central PMCID: PMC8036021.

[9] Martins CF, Serras I, Santos AV, Costa Braga A. Pentalogy of Cantrell: The complete spectrum. BMJ Case Rep. 2014;2014:bcr2014207556. Doi: 10.1136/bcr-2014-207556 PubMed PMID: 25378274; PubMed Central PMCID: PMC4225303.

[10] Toyama WM. Combined congenital defects of the anterior abdominal wall, sternum, diaphragm, pericardium, and heart: A case report and review of the syndrome. Pediatrics. 1972;50(5):778-92. PubMed PMID: 4263752.

[11] Mejías EM, Carrión E, Sparman A, Juliana A. What is pentalogy of Cantrell? Cardiol Young. 2024;1-3. Doi: 10.1017/S1047951124025794 PubMed PMID: 39358849.

[12] Naburi H, Assenga E, Patel S, Massawe A, Manji K. Class II pentalogy of Cantrell. BMC Res Notes. 2015;8:318. Doi: 10.1186/s13104-015-1293-7 PubMed PMID: 26219662; PubMed Central PMCID: PMC4517504.

[13] Berarducci J, Serrano-Roman JI, Armenta-Moreno JI, Cano-Zarate R, Espinola-Zavaleta N. Pentalogy of Cantrell. CJC Pediatr Congenit Heart Dis. 2022;1(4):198-99. Doi: 10.1016/j.cjcpc.2022.06.003 PubMed PMID: 37969932; PubMed Central PMCID: PMC10642150.

[14] Albrecht MEM, Sevillano Filho R, Araujo Júnior E. Late prenatal diagnosis of complete pentalogy of Cantrell. Ceska Gynekol. 2025;90(3):226-30. Doi: 10.48095/cccg2025226 PubMed PMID: 40663450.

[15] Zhu Y, Elhoff J, Rothman A, Kaushal S, Ciccolo ML. Surgical repair of tetralogy of fallot and a large congenital diaphragmatic hernia in a 16-week-old infant with pentalogy of cantrell and a large omphalocele: A case report. World J Pediatr Congenit Heart Surg. 2025;21. Epub ahead of print. Doi: 10.1177/21501351251375452 PubMed PMID: 41204094.

[16] Liao J, Huang H, Li X. Surgical treatment of neonatal Cantrell pentalogy: A case report and literature review. AME Case Rep. 2023;7:22. Doi: 10.21037/acr-23-14 PubMed PMID: 37492795; PubMed Central PMCID: PMC10364013.

[17] Hubbard R, Hayes S, Gillis H, Lindsey S, Malhotra P, Wani T, et al. Management challenges in an infant with pentalogy of cantrell, giant anterior encephalocele, and craniofacial anomalies: A case report. AA Pract. 2018;11(9):238-40. Doi: 10.1213/XAA.0000000000000793 PubMed PMID: 29782331.

[18] Sulistyowati R, Sensusiati AD. Radiological findings of partial expression pentalogy of Cantrell and other multiple congenital anomalies: A rare case report. Radiol Case Rep. 2022;17(9):3172-78. Doi: 10.1016/j.radcr.2022.05.083 PubMed PMID: 35801122; PubMed Central PMCID: PMC9253550.

[19] Meena RK, Meena V. Pentalogy of cantrell with single umbilical artery: A rare fetal anomaly. Int J Contemp Pediatr. 2017;4(1):280-82. Doi: 10.18203/2349-3291.ijcp20164620.

[20] Boyd JH, Lief S, Ansari AZ, Lichtenberg AB, De La Paz M, Patibandla S, et al. Pentalogy of Cantrell marked with ectopia cordis, gastroschisis, and cystic hygroma in the first trimester: A rare case. Cureus. 2024;16(9):e69360. Doi: 10.7759/cureus.69360 PubMed PMID: 39416577; PubMed Central PMCID: PMC11480700.

[21] Taee N, Goodarzi MF, Safdari M, Bajelan A. Pentalogy of Cantrell in full term neonate. AJP Rep. 2019;9(2):e144-e146. Doi: 10.1055/s-0039-1683936 PubMed PMID: 30972230; PubMed Central PMCID: PMC6456328.

[22] Aldeiri B, Roostalu U, Albertini A, Behnsen J, Wong J, Morabito A, et al. Abrogation of TGF-beta signalling in TAGLN expressing cells recapitulates Pentalogy of Cantrell in the mouse. Sci Rep. 2018;8(1):3658. Doi: 10.1038/s41598-018-21948-z PubMed PMID: 29483576; PubMed Central PMCID: PMC5826924.

[23] Mraihi F, Basly J, Mezni A, Ghali Z, Hafsi M, Chelli D. The pentalogy of Cantrell: A rare and challenging prenatal diagnosis. Int J Surg Case Rep. 2023;112:108941. Doi: 10.1016/j.ijscr.2023.108941 PubMed PMID: 37879290; PubMed Central PMCID: PMC10667751.

[24] Liang X, Ye L. Analysis of prenatal ultrasound manifestations in 15 cases of Cantrell Syndrome. Sichuan Da Xue Xue Bao Yi Xue Ban. 2024;55(3):749-55. Doi: 10.12182/20240560208 PubMed PMID: 38948286; PubMed Central PMCID: PMC11211774.

[25] Carmi R, Boughman JA. Pentalogy of Cantrell and associated midline anomalies: A possible ventral midline developmental field. Am J Med Genet. 1992;42(1):90-95. Doi: 10.1002/ajmg.1320420118 PubMed PMID: 1308371.

[26] Mărginean C, Mărginean CO, Gozar L, Meliţ LE, Suciu H, Gozar H, et al. Cantrell Syndrome-a rare complex congenital anomaly: A case report and literature review. Front Pediatr. 2018;6:201. Doi: 10.3389/fped.2018.00201 PubMed PMID: 30065917; PubMed Central PMCID: PMC6056637.

[27] Zeng D, Chen J, Liu J, Luo S. Pentalogy of Cantrell: A case report from China. Asian J Surg. 2024;S1015-9584(24)01463-5. Doi: 10.1016/j.asjsur.2024.07.093 PubMed PMID: 39034249.

[28] Ku L, Ma X. Cantrell syndrome. Eur Heart J. 2023;44(5):429. Doi: 10.1093/eurheartj/ehac563 PubMed PMID: 36210783.

[29] Steele J, Stewart RD, Pettersson G, Attia T, Komarlu R, Edwards T. Pentalogy of Cantrell: A view from fetus to operation with multimodality imaging. World J Pediatr Congenit Heart Surg. 2016;7(4):525-26. Doi: 10.1177/2150135116651096 PubMed PMID: 27358306.

[30] Benomar A, Bigras JL, Lapierre C. Pentalogy of Cantrell with complete ectopia cordis. Radiol Cardiothorac Imaging. 2025;7(3):e250030. Doi: 10.1148/ryct.250030 PubMed PMID: 40310286.

[31] Wang D, Zheng B, Zhai B, Mo J, Yang K, Huo Y. Clinical and ultrasound findings of pentalogy of Cantrell in a newborn: A case report. Front Pediatr. 2022;10:998495. Doi: 10.3389/fped.2022.998495 PubMed PMID: 36458139; PubMed Central PMCID: PMC9705726.

[32] Reyes Caldelas MA, de la Mora Cervantes R, Vargas Hernández SI. Cantrell's pentalogy❾- Angiographic CT evaluation. J Cardiovasc Comput Tomogr. 2020;14(5):e42-e43. Doi: 10.1016/j.jcct.2019.01.003 PubMed PMID: 30723002.
[33] Victoria T, Andronikou S, Bowen D, Laje P, Weiss DA, Johnson AM, et al. Fetal anterior abdominal wall defects: Prenatal imaging by magnetic resonance imaging. Pediatr Radiol. 2018;48(4):499-512. Doi: 10.1007/s00247-017-3914-x PubMed PMID: 29550866.
[34] Zani-Ruttenstock E, Zani A, Honjo O, Chiu P. Pentalogy of Cantrell: is echocardiography sufficient in the neonatal period? Eur J Pediatr Surg Rep. 2017;5(1):e9-e11. Doi: 10.1055/s-0037-1599794 PubMed PMID: 28344916; PubMed Central PMCID: PMC5363335.
[35] Peng W, Li L, Yang R, Xie H, Lan Y. Prenatal ultrasound diagnosis of pentalogy of Cantrell: A case report. Medicine (Baltimore). 2025;104(32):e43674. Doi: 10.1097/MD.0000000000043674 PubMed PMID: 40797417; PubMed Central PMCID: PMC12338190.
[36] Türkçapar AF, Sargın Oruc A, Öksüzoglu A, Danışman N. Diagnosis of pentalogy of cantrell in the first trimester using transvaginal sonography and color Doppler. Case Rep Obstet Gynecol. 2015;2015:179298. Doi: 10.1155/2015/179298 PubMed PMID: 25802780; PubMed Central PMCID: PMC4352946.
[37] Araujo Júnior E, Zanforlin Filho SM, Guimarães Filho HA, Pires CR, Nardozza LMM, Moron AF. Diagnosis of Pentalogy of Cantrell by three-dimensional ultrasound in third trimester of pregnancy. A case report. Fetal Diagn Ther. 2006;21(6):544-47. Doi: 10.1159/000095670 PubMed PMID: 16969012.
[38] Naren Satya SM, Mayilvaganan KR, Prathyusha IS, Gautam MS, Raidu D, Amogh VN. A Recurrent case of Pentalogy of Cantrell: A rare case with sonological findings and review of literature. Pol J Radiol. 2017;82:28-31. Doi: 10.12659/PJR.900086 PubMed PMID: 28203309; PubMed Central PMCID: PMC5267621.
[39] Madi JM, Festugatto JR, Rizzon M, Agostini AP, Araújo BF de, Garcia RMR. Ectopia Cordis Associated with Pentalogy of Cantrell-A case report. Rev Bras Ginecol E Obstet Rev Fed Bras Soc Ginecol E Obstet. 2019;41(5):352-56. Doi: 10.1055/s-0039-1679878 PubMed PMID: 30912088.
[40] Grigore M, Furnica C, Esanu I, Gafitanu D. Pentalogy of Cantrell associated with unilateral anophthalmia: Case report and literature review. Medicine (Baltimore). 2018;97(31):e11511. Doi: 10.1097/MD.0000000000011511 PubMed PMID: 30075516; PubMed Central PMCID: PMC6081174.
[41] Maheshwari V, Sahoo M. Anesthesia for Pentalogy of Cantrell with surgical repair of tetralogy of fallot along with absent diaphragm: A case study. Ann Card Anaesth. 2025;28(2):181-83. Doi: 10.4103/aca.aca_213_24 PubMed PMID: 40237667; PubMed Central PMCID: PMC12058065.
[42] Karaçelik M, Oral A, Bilen Ç, Polatdemir K. Multidisciplinary treatment for a patient with Pentalogy of Cantrell in the COVID-19 pandemic. Turk Gogus Kalp Damar Cerrahisi Derg. 2024;32(2):228-31. Doi: 10.5606/tgkdc.dergisi.2024.24969 PubMed PMID: 38933310; PubMed Central PMCID: PMC11197420.
[43] Araujo Júnior E, Carrilho MC, Toneto BR, Guilhen JCS. Pentalogy of Cantrell: Prenatal diagnosis, delivery, and immediate postnatal surgical repair. J Neonatal Surg. 2017;6(2):32. Doi: 10.21699/jns.v6i2.503 PubMed PMID: 28770129; PubMed Central PMCID: PMC5538598.
[44] Zhang X, Xing Q, Sun J, Hou X, Kuang M, Zhang G. Surgical treatment and outcomes of pentalogy of Cantrell in eight patients. J Pediatr Surg. 2014;49(8):1335-40. Doi: 10.1016/j.jpedsurg.2014.06.003 PubMed PMID: 25092102.
[45] Basarahalli MR, Alluri D, Kini S, Sapre A. Successful surgical repair of complete pentalogy of Cantrell. J Indian Assoc Pediatr Surg. 2024;29(6):630-33. Doi: 10.4103/jiaps.jiaps_108_24 PubMed PMID: 39691924; PubMed Central PMCID: PMC11649046.
[46] Divkovic D, Kvolik S, Sipl M, Sego K, Puseljic S, Rakipovic-Stojanovic A, et al. A successful early gore-tex reconstruction of an abdominal wall defect in a neonate with Cantrell pentalogy: A case report and literature review. Clin Case Rep. 2015;3(1):19-23. Doi: 10.1002/ccr3.137 PubMed PMID: 25678967; PubMed Central PMCID: PMC4317205.
[47] Ugwu JO, Etukokwu KC, Modekwe VI, Ekwunife HO, Orjiakor SC, Ilokanuno CN, et al. Pentalogy of Cantrell: reconstructive challenges of two complete cases in a resource-constrained setting. J Indian Assoc Pediatr Surg. 2021;26(4):259-61. Doi: 10.4103/jiaps.JIAPS_127_20 PubMed PMID: 34385772; PubMed Central PMCID: PMC8323571.
[48] Alanís-Naranjo JM, De la Cruz-Pelayo J, de la Mora-Cervantes R, Gamboa-Abundis OA, Meave-González A, Cruz-Marmolejo MÁ. Pentalogy of Cantrell: Comprehensive Multimodal Imaging Evaluation in an Adult With Ectopia Cordis. JACC Case Rep. 2026;31(11):106922. Doi: 10.1016/j.jaccas.2026.106922. PMID: 41665586; PMCID: PMC13008523.
[49] Engels AC, Debeer A, Russo FM, Aertsen M, Aerts K, Miserez M, et al. Pericardio-amniotic shunting for incomplete pentalogy of Cantrell. Fetal Diagn Ther. 2017;41(2):152-56. Doi: 10.1159/000457122 PubMed PMID: 28196368.
[50] Lopez-Rincon RM, Plaza-Lloret M, Munoz-Monaco G, Mavarez AC. Fascinating image of pentalogy of Cantrell: Navigating challenges for optimal anesthesia care. Anesthesiology. 2024;140(5):1016-17. Doi: 10.1097/ALN.0000000000004877 PubMed PMID: 38427825.
[51] Costa JV da S, Melonio CEC, Vieira CB, Oliveira CMB de, Leal P da C, Servín ETN, et al. Anesthesia for surgical repair of the pentalogy of Cantrell: Case report. Braz J Anesthesiol. 2019;69(3):322-25. Doi: 10.1016/j.bjan.2018.09.002 PubMed PMID: 30470449; PubMed Central PMCID: PMC9391903.
[52] Martadiansyah A, Bernolian N, Mirani P, Lestari PM, Nugraha A, Stevanny B. Complex management of ectopia cordis complicated by pentalogy of cantrell: Report of two cases and review of current evidence. Int J Surg Case Rep. 2025;131:111353. Doi: 10.1016/j.ijscr.2025.111353 PubMed PMID: 40286694; PubMed Central PMCID: PMC12059706.
[53] van Hoorn JHL, Moonen RMJ, Huysentruyt CJR, van Heurn LWE, Offermans JPM, Mulder ALMT. Pentalogy of Cantrell: Two patients and a review to determine prognostic factors for optimal approach. Eur J Pediatr. 2008;167(1):29-35. Doi: 10.1007/s00431-007-0578-9 PubMed PMID: 17674044; PubMed Central PMCID: PMC2668557.

PARTICULARS OF CONTRIBUTORS:
1. Junior Resident, Department of General Surgery, Jawaharlal Nehru Medical College, Datta Meghe Institute of Higher Education and Research, Wardha, Maharashtra, India.
2. Professor, Department of General Surgery, Jawaharlal Nehru Medical College, Datta Meghe Institute of Higher Education and Research, Wardha, Maharashtra, India.
3. Associate Professor, Department of General Surgery, Jawaharlal Nehru Medical College, Datta Meghe Institute of Higher Education and Research, Wardha, Maharashtra, India.
4. Junior Resident, Department of General Surgery, Jawaharlal Nehru Medical College, Datta Meghe Institute of Higher Education and Research, Wardha, Maharashtra, India.
5. Undergraduate Student, Department of Medicine, Jawaharlal Nehru Medical College, Datta Meghe Institute of Higher Education and Research, Wardha, Maharashtra, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Shailabh,
Junior Resident, Department of General Surgery, Jawaharlal Nehru Medical College, Datta Meghe Institute of Higher Education and Research, Wardha-442107, Maharashtra, India.
E-mail: shailabh.bhadra@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Feb 14, 2026
- Manual Googling: Jun 13, 2026
- iThenticate Software: Jun 15, 2026 (2%)

ETYMOLOGY: Author Origin

EMENDATIONS: 6

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? No
- For any images presented appropriate consent has been obtained from the subjects. NA

Date of Submission: **Jan 28, 2026**
Date of Peer Review: **Mar 06, 2026**
Date of Acceptance: **Jun 17, 2026**
Date of Publishing: **Oct 01, 2026**