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Transition of Care of Pediatric Surgical Patients

The transition from pediatric to adult healthcare represents a critical and complex process, especially for surgical patients who have lived with congenital anomalies, chronic diseases, or post-operative conditions that require lifelong management. As advances in pediatric surgery, neonatal resuscitation, and perioperative care have significantly improved survival outcomes, many children with once-lethal conditions now reach adulthood. However, this increased longevity has created a new challenge: ensuring these patients continue to receive high-quality, condition-specific care within adult healthcare systems that may not be equipped to handle the nuances of pediatric-onset disorders.

Transition of care is not a singular moment but a multifaceted, developmental process that unfolds over time. It involves preparing adolescents and their families for the eventual transfer of clinical responsibility to adult healthcare teams. This shift must address not only the medical needs of the patient but also psychosocial, educational, and vocational aspects. Transition planning should promote autonomy, empower self-advocacy, and ensure continuity of care. Despite recognition of its importance, many pediatric surgical specialties still lack formal, structured transition programs, leading to gaps in care, delayed referrals, and suboptimal long-term outcomes.

One of the most pressing issues is the scarcity of adult providers trained or willing to manage patients with pediatric surgical histories. Many adult surgeons are unfamiliar with the long-term sequelae of congenital conditions or childhood interventions. For instance, patients who underwent correction for anorectal malformations or Hirschsprung disease in infancy may face lifelong bowel dysfunction, incontinence, or complications related to sexual and urological function. Similarly, those treated for esophageal atresia, spina bifida, or congenital cardiac disease often require surveillance for late-onset complications that fall outside the typical scope of adult care. Without appropriate transition, these individuals may disengage from follow-up until they present emergently—by which time their conditions may have progressed, sometimes irreversibly.

Pediatric surgical teams often continue care well into the patient's twenties, in part because of the lack of clear pathways or designated receiving adult services. While this continuity can offer reassurance, it creates ethical and logistical tensions. Pediatric providers must weigh their commitment to individual patients against the broader duty to care for new pediatric cases. The balance between beneficence—ensuring the best outcomes for the individual—and justice—fair distribution of finite resources—is delicate. The absence of transition policies also increases the risk of violating patient autonomy, particularly when

adolescents are not included in decisions about their future care or are unprepared to assume responsibility for managing their health.

Surveys of surgical and medical professionals confirm widespread recognition of these challenges. A substantial majority acknowledge the benefits of structured transition models and advocate for standardized protocols. Yet implementation remains patchy, hindered by a lack of resources, training, and institutional support. Many health systems do not mandate transition policies, and even where guidelines exist, they are often not tailored to the surgical population. The result is a fragmented, inconsistent experience for patients and families.

Disease-specific approaches have emerged in response to this gap. In chronic intestinal failure, where patients rely on home parenteral nutrition, international experts have developed consensus protocols grounded in clinical experience and theory. These protocols emphasize early initiation of transition planning—ideally 1–2 years before the anticipated transfer—and include joint clinics staffed by pediatric and adult teams, transitional care coordinators, and standardized readiness assessments. Tailored interventions, such as education on catheter care and emergency management, address the unique risks these patients face. Central to the protocol's success is a nurse specialist embedded in both the pediatric and adult services, serving as a bridge and consistent presence throughout the transition.

Similarly, research on adolescents with inflammatory bowel disease underscores the importance of maturity-based rather than age-based transitions. Clinical data reveal that early-onset Crohn's disease is often more aggressive than its adult-onset counterpart, associated with higher surgical risk and greater likelihood of biologic therapy. This phenotype, combined with the psychosocial burden of a chronic illness during adolescence, demands a nuanced, individualized transition plan. A failure to manage the transition effectively can lead to disease flares, reduced medication adherence, and poor health-related quality of life.

In colorectal surgery, conditions such as anorectal malformations and Hirschsprung disease present particular transition challenges. Patients often develop a dependency on their pediatric teams and express reluctance to engage with unfamiliar adult services. Many lack understanding of their own medical history or the implications of their condition in adulthood. Concurrently, adult providers frequently report insufficient knowledge of pediatric colorectal pathologies and express discomfort managing their sequelae. In the absence of structured joint clinics or shared protocols, these patients face a heightened risk of discontinuity of care.

A systematic review of transitional care in colorectal surgery identified three overarching categories of barriers: patient-related (e.g., limited knowledge, psychological readiness), provider-related (e.g., inadequate training, lack of communication), and system-level (e.g., absence of joint clinics, lack of standardized pathways). Solutions to these challenges include fostering autonomy and self-efficacy among patients through education, training

adult clinicians in pediatric-onset conditions, and establishing multidisciplinary transition clinics. The creation of condition-specific guidelines is also critical, as generic transition frameworks often fail to capture the complexities of surgical diseases.

From an ethical standpoint, transition of care must be grounded in respect for autonomy, beneficence, non-maleficence, and justice. Transition planning should begin early, with adolescents actively participating in decisions about their future care. Patients must be given the tools to develop health literacy, navigate insurance changes, and manage treatment regimens independently. Simultaneously, institutions have a responsibility to ensure that adult providers are prepared and resourced to care for this unique population. Without such infrastructure, patients may face gaps in care or even harm.

One underappreciated dimension of transition is the role of genetics and personalized medicine. In inflammatory bowel disease, for example, studies have demonstrated significant associations between specific genetic polymorphisms and disease severity, location, and likelihood of surgery. Knowledge of a patient's genetic risk profile could inform timing and content of transition planning, particularly for those at greater risk of complications. It may also guide selection of adult providers with expertise in managing high-risk cases.

Despite growing awareness and international efforts to define best practices, many health systems remain unprepared to implement comprehensive transition models. Barriers include limited personnel, lack of training, and inadequate reimbursement for transition-related services. Moreover, the absence of a formal transition policy at the national or institutional level means that many clinicians continue to rely on informal, ad hoc arrangements. This variability exacerbates disparities in care, particularly for patients from underrepresented backgrounds or those with complex social needs.

Effective transition of care for pediatric surgical patients requires more than institutional commitment; it demands cultural change across pediatric and adult disciplines. Adult providers must be equipped not only with clinical knowledge but also with an appreciation of the developmental and emotional needs of patients who are emerging from a highly supportive pediatric environment. Conversely, pediatric teams must be willing to relinquish care at the appropriate time and to support patients and families through the uncertainty of change.

At its core, successful transition is about preserving the continuity, quality, and humanity of care. The transition process must respect the history of each patient's journey while preparing them for the road ahead. It requires coordination, communication, and above all, a shared commitment to supporting young adults as they move forward with their lives.

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Syndromic Biliary Atresia

Syndromic biliary atresia represents a distinct minority of biliary atresia cases but poses challenges that differ from isolated disease. About ten percent of infants with biliary atresia present with splenic and laterality anomalies that define the biliary atresia splenic malformation spectrum. These patients frequently show polysplenia, double spleens, or rarely asplenia, in combination with features such as situs inversus, preduodenal portal vein, absent inferior vena cava, intestinal malrotation, and a broad range of cardiac anomalies. The presence of these abnormalities signals a developmental disturbance early in embryogenesis. Imaging and operative findings often show atypical vascular arrangements, single or preduodenal portal veins, and altered biliary anatomy. These anomalies complicate exposure and reconstruction but do not preclude success when handled with sound technique.

The syndrome begins with early onset of biliary obstruction. Several studies note that infants with splenic malformations tend to declare their disease sooner than non-syndromic infants, which drives them to surgery at a younger age. Pooled data confirm this pattern, showing that the age at Kasai operation is consistently lower by about ten to thirteen days among those in the syndromic group. The earlier disease onset appears to reflect an intrinsic pace of biliary injury rather than differences in referral patterns. In some cases, early stools may still contain bile, misleading clinicians and briefly delaying diagnosis, but the underlying process remains aggressive.

Early surgery is a clear advantage and remains the most important modifiable factor in management. Across the collected series, successful Kasai reconstruction within the first sixty days of life offers the best chance of long-term native liver survival. Even in syndromic disease, timely surgery supports meaningful long-term outcomes. Operative illustrations in the reports show absence of gallbladder or biliary tree, a fibrotic portal plate, and anomalous portal venous structures. These views remind surgeons that dissection planes may differ, and exposure must be deliberate. The dissection should aim to maximize the surface of exposed microscopic ductules while remaining alert for aberrant portal vein

courses. A single portal vein or preduodenal configuration demands precise management to avoid injury, particularly during the portoenterostomy.

Historically, syndromic biliary atresia carried a reputation for poor outcomes. Several earlier reports suggested higher rates of postoperative complications, including cholangitis, persistent jaundice, and portal hypertension with variceal bleeding. Some believed that prognosis was impaired because the anomalies made the Kasai operation more complex, particularly in the presence of situs inversus or severe vascular rearrangement. Others emphasized that associated cardiac defects may influence outcomes independently. Yet the cumulative evidence from the seven reviewed studies presents a more balanced picture.

When outcomes across more than two thousand patients were pooled, no significant difference was found between syndromic and non-syndromic groups for jaundice clearance or native liver survival. Clearance of jaundice showed overlapping performance between groups, with odds ratios near unity and low heterogeneity. Native liver survival at follow-up periods ranging from one to twenty years also showed no clear divergence between cohorts. Differences noted in some earlier work appear to be a function of small cohorts, variable definitions, and inconsistent follow-up durations. Once follow-up time is standardized across subgroups, the survival curves between groups lose their earlier separation.

The operative challenges remain real, yet they do not reliably predict failure. Increasing surgical experience and improved imaging to define vascular anomalies help to equalize outcomes. Preoperative planning with high-quality ultrasound, magnetic resonance imaging, and attention to the portal venous trajectory is essential. Surgeons should anticipate aberrant structures and approach the hilum with caution. Despite these challenges, the technical goals remain the same as for isolated disease: complete clearance of fibrotic tissue overlying the portal plate and an unobstructed, tension-free Roux limb.

The postoperative course follows a familiar pattern. Infants face risk of cholangitis, persistent jaundice, hepatopulmonary syndrome, and progressive portal hypertension. Cholangitis stands out as a key predictor of poor outcomes when recurrent or severe. Some centers describe improvement with early postoperative steroid therapy, noting better bilirubin profiles and increased rates of jaundice clearance within the first six months. Steroids are usually tapered within a few months. While their long-term influence remains debated, they may optimize early recovery in selected cases.

New data from long-term cohorts bring a surprising insight into the biology of syndromic disease. Studies analyzing biomarkers of liver fibrosis over more than three decades found that syndromic groups, both BASM and non-BASM variants, showed lower markers of fibrosis compared with isolated biliary atresia. Platelet counts were higher at the time of Kasai and at three-year follow-up, suggesting less splenic sequestration and milder portal hypertension. The AST-to-platelet index ratio was lower in syndromic infants compared with

isolated disease, and the varices prediction rule consistently showed a lower likelihood of significant varices. Endoscopic findings confirm this biologic signal. Clinically significant varices were less common in syndromic biliary atresia, with rates near four percent compared with more than twenty percent in isolated disease. This pattern indicates that, despite early onset of biliary obstruction, the long-term fibrotic trajectory may be milder in some forms of syndromic biliary atresia.

The mechanism behind this is not fully understood. Genetic studies point toward variants in laterality and ciliopathy pathways, including genes such as CFC1 and PKD1L1. Other associated pathways influence ciliary morphogenesis, bile duct development, and laterality specification. Abnormal embryologic signaling may produce the extrahepatic anomalies that define the syndrome and simultaneously influence liver fibrosis patterns. Some studies suggest environmental contributions, including maternal diabetes in selected cases. The heterogeneity of associated conditions, such as Cat-Eye syndrome, Kabuki syndrome, Hardikar syndrome, and a wide range of nonrandom congenital associations, supports the view that syndromic biliary atresia is not a single disease but a convergence of several developmental disorders that share a final pathway of biliary obstruction.

Understanding these mechanisms may eventually guide personalized strategies. For surgeons, though, the immediate implication is that syndromic anatomy does not always predict poor long-term liver health. A child with polysplenia or malrotation may still experience milder portal hypertension years after surgery compared with a child with isolated disease. The risk of requiring transplantation remains present, but some cohorts show durable native liver survival in both BASM and non-BASM syndromic groups comparable to isolated disease.

These findings do not imply complacency. Careful follow-up remains necessary because syndromic infants still display wide variability in postoperative stability. Even when jaundice clears early, subtle progression of fibrosis or late complications such as hepatopulmonary syndrome can occur. Surveillance strategies should include routine liver biochemistry, growth assessment, ultrasound with Doppler evaluation, endoscopy when clinically indicated, and consideration of biomarkers such as APRi and VPR to guide timing of intervention. Early management of complications can prevent avoidable morbidity.

While the literature shows no significant difference in survival, the limited number of controlled studies and small cohort sizes restrict firm conclusions. Many analyses rely on retrospective data, and definitions of jaundice clearance vary across publications. Time-to-event strategies like Kaplan-Meier or Cox regression are ideal but often under-reported. Future studies with larger, prospective designs may establish clearer prognostic signals and help distinguish which anatomic or genetic subgroups behave differently.

As current data stand, the key principles for surgeons treating syndromic biliary atresia remain consistent. Early diagnosis is essential. Early Kasai reconstruction within the first two months improves the chance of long-term survival, regardless of anatomic anomalies. Detailed preoperative imaging and deliberate intraoperative technique can overcome the

hurdles posed by atypical vascular or biliary anatomy. Postoperative care mirrors that of isolated biliary atresia, though syndromic patients may show different patterns of fibrosis and portal hypertension. Steroid-enhanced recovery in the early postoperative period may play a role for selected patients. Long-term follow-up should be individualized and attentive to baseline anomalies.

Syndromic biliary atresia is an important subgroup that challenges the surgeon in diagnosis, operative planning, and long-term care. Yet the collective evidence shows that, when treated early and skillfully, these infants can achieve outcomes similar to those with isolated disease. Variability persists, but syndromic anatomy alone should not dictate prognosis or limit the pursuit of full biliary reconstruction. With continued advances in imaging, genetics, and postoperative management, surgeons can approach syndromic biliary atresia with greater clarity, grounded in the knowledge that careful technique and early intervention remain the strongest determinants of success.

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Pyomyositis

Pyomyositis is a primary bacterial infection of skeletal muscle that leads to localized inflammation and frequently abscess formation. Once considered a disease largely confined to tropical climates, its growing recognition in temperate regions has reshaped epidemiologic assumptions and raised new clinical questions. Across recent observational studies, systematic reviews, and case series, pyomyositis emerges as a complex infection influenced by microbial, host, and environmental factors, and one that continues to challenge clinicians because of its variable presentation and potential for severe complications.

The historical characterization of pyomyositis as “tropical myositis” reflected early epidemiologic patterns, with reports from sub-Saharan Africa and other equatorial areas describing high burdens and notable contributions to surgical caseloads. A systematic review of global studies underscores this origin while confirming increasing reports from temperate regions, where previously the disease had been rare. This geographic shift is

echoed in pediatric studies from Europe and Japan, which document a rising number of cases over the past decades. The consistency of these reports argues that pyomyositis is no longer a tropical infection alone, but a globally relevant condition influenced by both environmental exposure and host susceptibility.

From a demographic perspective, pyomyositis affects children and adults of all ages, but many cohorts report a predominance of cases in the pediatric population. Several studies note a skew toward males, particularly in younger age groups, with one meta-analysis identifying males under 20 years as the most frequently affected demographic. In a decade-long pediatric study focusing on pelvic pyomyositis, two age peaks were identified: children under two years and adolescents, suggesting different exposure profiles or physiological vulnerabilities across developmental stages.

Trauma has long been recognized as a contributing factor, and multiple cohorts reinforce this association. In one pediatric pelvis-focused study, nearly a quarter of children reported recent trauma, and some had associated skin lesions or recent intramuscular injections. In a large extremity-focused series, trauma preceded symptom onset in nearly 70 percent of cases, highlighting the possibility that muscle microinjury enables bacterial seeding or growth .

Immunosuppression also plays a critical role. A meta-analysis evaluating factors associated with pyomyositis found strong correlations with HIV infection and advanced immunosuppression, with odds ratios as high as six for those with AIDS-defining illness. Other reported comorbidities include diabetes, hematologic malignancies, renal disease, and autoimmune conditions. Even among otherwise healthy children, concurrent viral or respiratory infections are not uncommon, raising the possibility that transient immune modulation may increase vulnerability to bacterial invasion.

Across studies, *Staphylococcus aureus* remains the dominant pathogen. Numerous pediatric cohorts describe *S. aureus* isolation in 30 to 90 percent of culture-positive cases, with both methicillin-sensitive and methicillin-resistant strains represented . Several systematic reviews similarly identify *S. aureus* as the principal organism, accounting for nearly four out of five cases in some analyses. In one pediatric case series from Somalia, cultures grew *S. aureus* in 69 percent of patients, reinforcing the organism’s central role across continents and healthcare environments.

Despite the predominance of *S. aureus*, less common pathogens are increasingly recognized. A detailed pediatric case report described pyomyositis caused by *Streptococcus pneumoniae*, a rare but important cause of invasive muscle infection. The report highlighted severe systemic inflammation, rapid progression, and abscess formation involving multiple pelvic muscles . Such cases emphasize that while empirical therapy should prioritize staphylococcal coverage, clinicians must remain alert to atypical causes, particularly in severe or refractory disease.

The presentation of pyomyositis is notoriously variable. Early symptoms are non-specific

and may include low-grade fever, localized muscle pain, and subtle functional limitations. These features often mimic more common conditions such as muscle strain or transient synovitis, contributing to diagnostic delays. In the pediatric pelvis-focused study, pain, functional limitation, and fever were the most frequent presenting symptoms, yet diagnostic delays averaged five days from symptom onset and were even longer in younger children, who tend to present with irritability rather than localized pain.

Across multiple reports, the muscles of the pelvis and thigh are the most frequently involved sites, including the iliopsoas, obturator muscles, and gluteal groups. A Japanese 32-year institutional review demonstrated that more than 90 percent of pyomyositis cases involved muscles adjacent to the hip joint, reinforcing this anatomical pattern in temperate climates as well. Studies focusing on extremity involvement note similar trends, with lower extremity muscles affected more often than upper extremity groups.

Laboratory markers such as C-reactive protein, erythrocyte sedimentation rate, and white blood cell count are usually elevated but lack diagnostic specificity. In pediatric patients, C-reactive protein tends to be significantly higher in older children, while very young infants may show subtler laboratory abnormalities despite significant infection .

Imaging is the cornerstone of diagnosis. Magnetic resonance imaging (MRI) consistently emerges as the most reliable modality, capable of detecting intramuscular edema, abscesses, and involvement of adjacent structures with high sensitivity. In the pelvic pyomyositis study, MRI detected abnormalities in every case, even when clinical and laboratory findings were inconclusive. The addition of diffusion-weighted imaging further enhanced detection of deep or early infection .

Systematic reviews confirm MRI as the diagnostic gold standard, though ultrasound remains a useful initial tool due to its accessibility, particularly for identifying fluid collections suitable for aspiration . Plain radiographs are generally unremarkable early in disease but can help rule out alternative diagnoses. In acute musculoskeletal infection more broadly, combined clinical, laboratory, and imaging findings are considered essential because no single test offers definitive sensitivity or specificity .

Pyomyositis has been traditionally divided into three stages: invasive, suppurative, and late or septic. Early disease can remain subtle for days before progressing to abscess formation. In the extremity-focused case series, most children presented after an average of two weeks of symptoms, reflecting the difficulty of recognizing early-stage disease and the rapid progression to abscess formation in many cases.

Antibiotic therapy and drainage remain the primary treatments. Empiric therapy typically includes coverage for methicillin-sensitive and methicillin-resistant *S. aureus*, with broad-spectrum regimens initiated when necessary. In a reported case of *S. pneumoniae* pyomyositis, initial broad coverage was appropriately narrowed once the organism was identified, but the child still required six weeks of combined intravenous and oral therapy, highlighting the prolonged courses often required for complete resolution.

Surgical drainage is often necessary, particularly in the suppurative stage. A systematic review found that medical therapy alone was successful in approximately 40 percent of cases but that surgical or percutaneous drainage was required in the remainder, especially when abscess size was significant or when clinical deterioration occurred. Larger inflammatory markers and more severe symptoms at presentation predicted the need for operative intervention.

In the large Somali pediatric series, all children underwent surgical debridement, with nearly universal recovery. Only a small subset developed complications such as osteomyelitis, demonstrating that with timely intervention, outcomes can be favorable even in resource-limited settings.

Complications across studies include osteomyelitis, septic arthritis, sepsis, and multifocal infection. Methicillin-resistant *S. aureus* has been identified as a predictor of more severe disease and higher complication rates. Pediatric reports also highlight the risk of multifocal infection, particularly in pelvic disease where adjacent joints and bones are frequently involved.

The emerging epidemiology of pyomyositis reflects broader changes in global health patterns. The increase in temperate-region cases reported in recent pediatric and adult cohorts suggests improved recognition, greater use of advanced imaging, or possibly true shifts in disease distribution. The Japanese 32-year institutional review documented a notable rise in cases in the past sixteen years compared to the previous sixteen, although the difference did not reach statistical significance. Nevertheless, this trend reinforces the need for heightened clinical suspicion, even in areas where pyomyositis was once considered rare.

Pyomyositis is a globally relevant infection marked by diverse presentations, evolving epidemiology, and the continued dominance of *Staphylococcus aureus* as the primary pathogen. Though early symptoms can be subtle, timely recognition is essential to prevent complications. MRI stands as the most sensitive diagnostic tool, and management typically requires a combination of targeted antimicrobial therapy and drainage of abscesses when present. Recent studies from Europe, Asia, and Africa highlight the infection's rising incidence in temperate regions and underline the importance of including pyomyositis in the differential diagnosis of musculoskeletal infections, particularly in children presenting with fever, pain, and functional limitation. Continued research is needed to clarify pathogenesis, refine diagnostic pathways, and optimize treatment strategies for this challenging and often under-recognized disease.

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